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# DIAGNOSTICS OF THE FUNDUS OCULI

BY

EDWARD L. OATMAN, M. D.

SURGEON MANHATTAN EYE, EAR AND THROAT HOSPITAL AND BROOKLYN EYE AND EAR HOSPITAL,  
NEW YORK, CONSULTING OPHTHALMIC SURGEON NYACK HOSPITAL  
AND ST. MARY'S HOSPITAL, WATERBURY, CONN.

COMPRISING  
ONE VOLUME OF TEXT  
*WITH TWO HUNDRED AND THIRTY-FOUR ILLUSTRATIONS*  
*AND FOUR COLORED PLATES*  
AND  
TWO PORTFOLIOS  
*CONTAINING SEVENTY-NINE STEREOGRAMS AND*  
*EIGHT DIAGNOSTIC CARDS*

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1920

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## PREFACE.

In the *Diagnostics of the Fundus Oculi*, the author seeks to present his subject in a manner serviceable to the ophthalmologist, the neurologist and the general practitioner, as well as to the teacher and the student. It is hoped that the ophthalmologist may find in the text, with its accompanying figures, some features that will meet with his approbation, and that the stereographic reproductions of the fundus will recall to his mind cases encountered in his own practice. There are many diseased conditions of the fundus with which the general practitioner should be familiar, and it is believed that the stereograms accompanying this volume will aid him greatly in their recognition. They are all reproductions of actual cases and in each instance the fundus conditions have been depicted with every effort possible toward accuracy. The rarer examples, such as those of maculo-cerebral degeneration and retinitis punctata albescens, are hardly necessary in his ordinary practice, but it would seem highly desirable and almost essential that he should be able to call to his aid in diagnosis such common fundus pictures as those presented by choked disc, albuminuric retinitis and optic atrophy, with others less common, perhaps, as his knowledge of fundus diseases and his facility in the use of the ophthalmoscope increase. The teacher will, it is hoped, find the stereograms of advantage as substitutes, when necessary, for actual cases in his clinical lectures, and they can hardly fail to be of a similar service to the student of ophthalmology. It is not without some misgivings that a work of this character is presented to the medical profession. Many extensive works on diseases of the fundus have preceded it, as have also treatises more profound upon its pathology. It has been the endeavor of the author to combine both these features in a work sufficiently condensed for agreeable study. The collection of cases and specimens has extended over a period of a number of years and, in his efforts in this direction, the author has to thank many of his colleagues who have so kindly given their aid and to whom reference is made, it is hoped in all instances, in the text. In the preparation of the pathological specimens, his thanks are especially due to Dr. James Ewing and Dr. Henry T. Brooks, and he has also to thank Dr. Alfred Braun for his kindly criticism of the color tints, as well as Dr. H. W. Wootton for suggestions in regard to the text.

E. L. OATMAN.

40 East 41st Street.



## CONTENT OF PART I.

### CHAPTER I.

#### EXAMINATION OF THE EYE.

|                                     | PAGE |
|-------------------------------------|------|
| THE OPHTHALMOSCOPE .....            | 1    |
| USE OF THE OPHTHALMOSCOPE:          |      |
| Direct Method .....                 | 3    |
| Indirect Method .....               | 4    |
| FOCAL OR OBLIQUE ILLUMINATION ..... | 5    |
| TRANSILLUMINATION .....             | 7    |
| DIGITAL TONOMETRY .....             | 9    |
| INSTRUMENTAL TONOMETRY .....        | 10   |

### CHAPTER II.

#### THE NORMAL FUNDUS.

|                                       |    |
|---------------------------------------|----|
| BLOND FUNDUS .....                    | 11 |
| SCLERA .....                          | 11 |
| SUPRACHOROIDEA .....                  | 11 |
| CHOROID .....                         | 11 |
| RETINA .....                          | 13 |
| RETINAL REFLEXES—NEGROID FUNDUS ..... | 14 |
| THE RETINAL VESSELS .....             | 15 |
| PULSE PHENOMENA .....                 | 16 |
| THE OPTIC NERVE .....                 | 17 |
| Anatomy .....                         | 18 |

### CHAPTER III.

#### CONGENITAL ANOMALIES.

|                                           |    |
|-------------------------------------------|----|
| COLOBOMA OF THE CHOROID .....             | 20 |
| COLOBOMA OF CHOROID AND OPTIC NERVE ..... | 21 |
| Pathogenesis .....                        | 22 |

|                                                         | PAGE |
|---------------------------------------------------------|------|
| COLOBOMA OF THE MACULA .....                            | 23   |
| Diagnosis .....                                         | 24   |
| COLOBOMA OF OPTIC NERVE AND NERVE SHEATH .....          | 24   |
| Diagnosis .....                                         | 26   |
| INFERIOR OPTIC CRESCENT (FUCHS' COLOBOMA) .....         | 26   |
| OPAQUE NERVE FIBERS (MEDULLATED NERVE FIBERS) .....     | 27   |
| Diagnosis .....                                         | 29   |
| PREPAPILLARY RETINAL ARTERY .....                       | 29   |
| Diagnosis .....                                         | 30   |
| PSEUDONEURITIS .....                                    | 30   |
| PERSISTENT HYALOID ARTERY—PERSISTENT CANAL OF CLOQUET.. | 32   |
| Diagnosis .....                                         | 33   |
| ALBINISM .....                                          | 33   |
| Diagnosis .....                                         | 34   |
| PUNCTATE CONDITION OF THE FUNDUS (GUNN'S DOTS) .....    | 35   |
| Diagnosis .....                                         | 36   |

## CHAPTER IV.

## AFFECTIONS OF THE CHOROID.

|                                                           |    |
|-----------------------------------------------------------|----|
| EXUDATIVE CHOROIDITIS .....                               | 37 |
| Pathology .....                                           | 37 |
| CHOROIDITIS DISSEMINATA, EXUDATIVE STAGE .....            | 37 |
| Diagnosis .....                                           | 39 |
| CHOROIDITIS DISSEMINATA, EXUDATIVE STAGE .....            | 40 |
| Symptoms .....                                            | 41 |
| Diagnosis .....                                           | 41 |
| CENTRAL EXUDATIVE CHOROIDITIS, EXUDATIVE STAGE .....      | 42 |
| Diagnosis .....                                           | 43 |
| CHOROIDITIS DISSEMINATA, CICATRICIAL STAGE .....          | 43 |
| Repair .....                                              | 44 |
| Diagnosis .....                                           | 45 |
| CENTRAL CHOROIDITIS, COMMENCING CICATRIZATION .....       | 45 |
| Diagnosis .....                                           | 46 |
| CHORIORETINITIS PIGMENTOSA; RETINITIC OPTIC ATROPHY ..... | 46 |
| Diagnosis .....                                           | 47 |

## CONTENTS.

vii

|                                                                                                    | PAGE |
|----------------------------------------------------------------------------------------------------|------|
| TUBERCULOSIS OF THE CHOROID .....                                                                  | 48   |
| Diagnosis .....                                                                                    | 51   |
| From Miliary Tuberculosis .....                                                                    | 51   |
| From Conglomerate Tubercle .....                                                                   | 51   |
| From Septic Retinitis .....                                                                        | 52   |
| From Focal Tuberculosis of the Choroid .....                                                       | 52   |
| DIFFUSE CHORIORETINITIS, ATROPHIC STAGE .....                                                      | 52   |
| VASCULAR DEGENERATION OF THE CHOROID .....                                                         | 54   |
| Diagnosis .....                                                                                    | 55   |
| ATROPHY OF THE CHOROID—ACQUIRED ALBINISM .....                                                     | 55   |
| Diagnosis .....                                                                                    | 57   |
| SENILE CHANGES IN THE FUNDUS—COLLOID EXCRESCENCES—MAC-<br>ULAR DEGENERATION—ARTERIOSCLEROSIS ..... | 57   |
| Diagnosis .....                                                                                    | 59   |
| RUPTURE OF THE CHOROID .....                                                                       | 59   |

## CHAPTER V.

### SCLEROSIS OF THE RETINAL VESSELS AND RETINAL HEMORRHAGE.

|                                                       |    |
|-------------------------------------------------------|----|
| DIFFUSE RETINAL ARTERIOSCLEROSIS, PRIMARY STAGE ..... | 62 |
| Etiology .....                                        | 63 |
| Diagnosis .....                                       | 64 |
| ADVANCED SCLEROSIS OF RETINAL VESSELS .....           | 65 |
| Diagnosis .....                                       | 66 |
| Morbid Anatomy .....                                  | 68 |
| ARTERIOSCLEROSIS AND OPTIC NEURITIS .....             | 69 |
| Diagnosis .....                                       | 69 |
| RETINAL HEMORRHAGE IN ARTERIOSCLEROSIS .....          | 70 |
| RETINAL HEMORRHAGE .....                              | 71 |
| Etiology of Retinal Hemorrhage in General .....       | 73 |
| Arteriosclerosis .....                                | 73 |
| Anemia .....                                          | 74 |
| Chlorosis .....                                       | 74 |
| Malaria .....                                         | 74 |
| Syphilis .....                                        | 74 |
| Stasis .....                                          | 74 |
| Natal Retinal Hemorrhage .....                        | 75 |



|                                           |            |
|-------------------------------------------|------------|
| PRERETINAL OR SUBHYALOID HEMORRHAGE ..... | PAGE<br>76 |
| INTRAVITREOUS HEMORRHAGE .....            | 77         |

## CHAPTER VI.

## ANGIOPATHIC RETINITIS.

|                                                   |     |
|---------------------------------------------------|-----|
| ANGIOPATHIC RETINITIS IN GENERAL .....            | 80  |
| ANGIOPATHIC ALBUMINURIC RETINITIS .....           | 81  |
| PRECURSORY STAGE .....                            | 81  |
| Diagnosis .....                                   | 82  |
| DEVELOPED ANGIOPATHIC ALBUMINURIC RETINITIS ..... | 82  |
| Symptoms .....                                    | 82  |
| History and Course .....                          | 84  |
| Pathology .....                                   | 85  |
| ACUTE ALBUMINURIC RETINITIS .....                 | 86  |
| Exudative Stage .....                             | 88  |
| Diagnosis .....                                   | 89  |
| Degenerative Stage .....                          | 90  |
| HEMORRHAGIC ALBUMINURIC RETINITIS .....           | 90  |
| ANGIOPATHIC ALBUMINURIC NEURORETINITIS .....      | 90  |
| Diagnosis .....                                   | 91  |
| Atrophic Stage, Retinitic Optic Atrophy .....     | 92  |
| ANGIOPATHIC DIABETIC RETINITIS .....              | 94  |
| CENTRAL PUNCTATE DIABETIC RETINITIS .....         | 94  |
| HEMORRHAGIC DIABETIC RETINITIS .....              | 95  |
| ANGIOPATHIC LEUKEMIC RETINITIS .....              | 96  |
| LEUKEMIC NEURORETINITIS .....                     | 96  |
| HEMORRHAGIC LEUKEMIC RETINITIS .....              | 98  |
| RETINITIS IN PERNICIOUS ANEMIA .....              | 98  |
| ANGIOPATHIC SYPHILITIC RETINITIS .....            | 100 |
| RETINITIS CIRCINATA .....                         | 101 |

## CHAPTER VII.

## CLOSURE OF THE RETINAL VESSELS.

|                                                                     | PAGE |
|---------------------------------------------------------------------|------|
| CLOSURE OF THE CENTRAL ARTERY .....                                 | 105  |
| Ophthalmoscopic Appearance .....                                    | 105  |
| Interpretation of the Ophthalmoscopic Picture .....                 | 106  |
| Foveal Red Spot .....                                               | 107  |
| Retinal Vessels .....                                               | 108  |
| Granular Current .....                                              | 111  |
| Hemorrhage .....                                                    | 111  |
| Etiology .....                                                      | 113  |
| Embolism .....                                                      | 113  |
| CLOSURE OF CENTRAL ARTERY WITH VISIBLE COLLATERAL CIRCULATION ..... | 115  |
| Etiology (continued)                                                |      |
| Angiospasm .....                                                    | 119  |
| Thrombosis .....                                                    | 122  |
| Arteriosclerosis .....                                              | 124  |
| Unusual Causes .....                                                | 125  |
| Anatomical Characters .....                                         | 126  |
| Clinical History .....                                              | 132  |
| Diagnosis .....                                                     | 133  |
| Occlusion Atrophy .....                                             | 133  |
| RETINAL ATROPHY FOLLOWING CLOSURE OF CENTRAL ARTERY ....            | 134  |
| Branch Artery Closure .....                                         | 134  |
| Diagnosis .....                                                     | 136  |
| CLOSURE OF THE CENTRAL RETINAL VEIN .....                           | 138  |
| Diagnosis .....                                                     | 140  |
| Etiology .....                                                      | 140  |
| OBSTRUCTION OF CENTRAL VEIN WITH NEURITIS .....                     | 142  |
| ANATOMICAL CHARACTERS OF CLOSURE OF CENTRAL VEIN .....              | 143  |
| Thrombosis .....                                                    | 147  |
| Clinical History .....                                              | 150  |
| Complications .....                                                 | 151  |
| CLOSURE OF VENOUS BRANCH .....                                      | 155  |
| POST-THROMBOTIC CHANGES .....                                       | 156  |
| OBSTRUCTION OF BOTH CENTRAL VESSELS .....                           | 156  |

## CHAPTER VIII.

## DETACHMENT OF THE RETINA.

|                                                    | PAGE |
|----------------------------------------------------|------|
| SEROUS DETACHMENT .....                            | 159  |
| INCIPIENT DETACHMENT .....                         | 160  |
| Diagnosis .....                                    | 160  |
| SYMPTOMS OF RETINAL DETACHMENT .....               | 161  |
| OPHTHALMOSCOPIC PICTURE OF RECENT DETACHMENT ..... | 163  |
| DIFFERENTIATION OF RETINAL DETACHMENT .....        | 164  |
| REATTACHMENT OF THE RETINA .....                   | 165  |
| OLD DETACHMENT OF THE RETINA .....                 | 166  |
| SYMPTOMS OF RETINAL DETACHMENT .....               | 167  |
| Prognosis .....                                    | 168  |
| Etiology .....                                     | 168  |
| Exudative Theory .....                             | 169  |
| Myopia, Mechanical Theory .....                    | 170  |
| Hemorrhagic Theory .....                           | 170  |
| Vitreous Detachment .....                          | 171  |
| Retraction Theory .....                            | 172  |
| Diffusion Theory .....                             | 173  |
| Hyperemic Theory .....                             | 174  |
| Iridocyclitis .....                                | 174  |
| Traumatism .....                                   | 175  |
| Unusual Causes .....                               | 176  |

## CHAPTER IX.

## DEGENERATIONS OF THE RETINA.

|                                           |     |
|-------------------------------------------|-----|
| RETINITIS PIGMENTOSA .....                | 179 |
| Ophthalmoscopic Picture .....             | 180 |
| Diagnosis .....                           | 181 |
| RETINITIS PIGMENTOSA, SINE PIGMENTO ..... | 181 |
| ATROPHIA GYRATA CHOROIDÆ ET RETINÆ .....  | 181 |
| COURSE OF RETINITIS PIGMENTOSA .....      | 183 |
| Complications .....                       | 183 |
| Etiology .....                            | 184 |
| Diagnosis .....                           | 185 |
| Anatomical Characters .....               | 186 |

## CONTENTS.

xi

|                                                      | PAGE    |
|------------------------------------------------------|---------|
| <b>RETINITIS PUNCTATA ALBESCENS</b> .....            | 189     |
| Ophthalmoscopic Picture .....                        | 190     |
| <b>MACULOCEREBRAL DEGENERATION</b> .....             | 191     |
| Ophthalmoscopic Picture .....                        | 193-194 |
| Pathologic Anatomy .....                             | 195     |
| Etiology .....                                       | 196     |
| Differentiation .....                                | 197     |
| From Hereditary Macular Anomaly .....                | 197     |
| From Retinitis Pigmentosa .....                      | 197     |
| From Amaurotic Family Idiocy .....                   | 198     |
| From Central Chorioretinitis .....                   | 199     |
| From Unclassified Cases .....                        | 199     |
| <b>AMAUROTIC FAMILY IDIOCY (TAY'S DISEASE)</b> ..... | 200     |
| Etiology .....                                       | 201     |
| Morbid Anatomy .....                                 | 202     |
| Ophthalmoscopic Picture .....                        | 203     |

## CHAPTER X.

### MISCELLANEOUS AFFECTIONS OF THE RETINA.

|                                                |         |
|------------------------------------------------|---------|
| <b>CYANOSIS RETINÆ</b> .....                   | 204     |
| Ophthalmoscopic Picture .....                  | 205     |
| Diagnosis .....                                | 205     |
| <b>TRAUMATIC EDEMA (COMMOTIO RETINÆ)</b> ..... | 205     |
| Ophthalmoscopic Picture .....                  | 207     |
| Diagnosis .....                                | 207     |
| <b>TOXIC AMAUROSIS AND AMBLYOPIA</b> .....     | 208     |
| From Wood Alcohol .....                        | 208     |
| From Chronic Alcoholism .....                  | 209     |
| From Tobacco .....                             | 209     |
| From Iodoform .....                            | 210     |
| From Quinine .....                             | 210     |
| From Salicylic Acid .....                      | 210     |
| From Less Frequent Causes .....                | 210-211 |
| <b>RETINITIS PROLIFERANS</b> .....             | 211     |
| Ophthalmoscopic Picture .....                  | 212     |

|                                                       | PAGE |
|-------------------------------------------------------|------|
| RETINITIS STRIATA AND ANGIOID STREAKS IN RETINA ..... | 213  |
| Ophthalmoscopic Picture .....                         | 214  |
| METASTATIC RETINITIS (ENDOGENOUS OPHTHALMITIS) .....  | 215  |
| Pathology .....                                       | 216  |
| SEPTIC RETINITIS OF ROTH .....                        | 217  |
| Pathology .....                                       | 218  |
| Diagnosis .....                                       | 218  |

## CHAPTER XI.

## OPTIC NEURITIS AND OPTIC ATROPHY.

|                                                                                 |         |
|---------------------------------------------------------------------------------|---------|
| OPTIC NEURITIS AND CHOKED DISC .....                                            | 220     |
| Ophthalmoscopic Picture .....                                                   | 222-224 |
| Diagnosis .....                                                                 | 222-224 |
| NEURITIS IN NASAL SINUS DISEASE .....                                           | 225     |
| Symptoms .....                                                                  | 226     |
| Ophthalmoscopic Picture .....                                                   | 227     |
| THE OPTIC NERVE IN CEREBRAL SINUS THROMBOSIS .....                              | 228     |
| Ophthalmoscopic Picture .....                                                   | 230     |
| OPTIC ATROPHY .....                                                             | 231     |
| Simple, in Tabes .....                                                          | 231     |
| Ophthalmoscopic Picture .....                                                   | 233     |
| In Multiple Sclerosis .....                                                     | 233     |
| Ophthalmoscopic Picture .....                                                   | 234     |
| In Other Diseases of the Nervous System .....                                   | 235-236 |
| NEURITIC OPTIC ATROPHY .....                                                    | 236     |
| Ophthalmoscopic Picture .....                                                   | 237     |
| DIFFERENTIATION OF SIMPLE, NEURITIC AND RETINITIC ATROPHY<br>AND GLAUCOMA ..... | 238     |
| COMPRESSION OF OPTIC NERVE IN ARTERIOSCLEROSIS .....                            | 239     |

## CHAPTER XII.

## MYOPIA.

|                    |     |
|--------------------|-----|
| AXIAL MYOPIA ..... | 244 |
| Etiology .....     | 244 |

## CONTENTS.

xiii

|                                               | PAGE |
|-----------------------------------------------|------|
| NON-PROGRESSIVE—OPHTHALMOSCOPIC PICTURE ..... | 246  |
| Anatomic Changes .....                        | 246  |
| PROGRESSIVE MYOPIA .....                      | 248  |
| Ophthalmoscopic Picture .....                 | 249  |
| CURVATURE MYOPIA .....                        | 249  |
| INDEX MYOPIA .....                            | 250  |

## CHAPTER XIII.

### GLAUCOMA.

|                                                                                                                                  |     |
|----------------------------------------------------------------------------------------------------------------------------------|-----|
| PRIMARY GLAUCOMA .....                                                                                                           | 251 |
| Prodromal Stage .....                                                                                                            | 251 |
| Acute Inflammatory Stage .....                                                                                                   | 251 |
| Habitus Glaucomatosus .....                                                                                                      | 253 |
| Glaucoma Absolutum .....                                                                                                         | 253 |
| Glaucoma Degenerativum .....                                                                                                     | 254 |
| Glaucoma Fulminans .....                                                                                                         | 254 |
| Chronic or Sub-acute Glaucoma .....                                                                                              | 254 |
| Simple Glaucoma .....                                                                                                            | 254 |
| Hemorrhagic Glaucoma .....                                                                                                       | 255 |
| OPHTHALMOSCOPIC PICTURE OF GLAUCOMA .....                                                                                        | 255 |
| Etiology of Primary Glaucoma .....                                                                                               | 257 |
| Secondary Glaucoma .....                                                                                                         | 260 |
| Morbid Anatomy .....                                                                                                             | 262 |
| Buphthalmus .....                                                                                                                | 264 |
| Ophthalmoscopic Picture and Microscopic Examination of<br>So-called Hemorrhagic Glaucoma in Advanced Arterio-<br>sclerosis ..... | 265 |
| DIAGNOSIS OF GLAUCOMA IN GENERAL .....                                                                                           | 266 |
| From Iridocyclitis and Iritis .....                                                                                              | 268 |

## CHAPTER XIV.

### INTRAOCULAR AND ORBITAL TUMORS.

|                                    |         |
|------------------------------------|---------|
| SARCOMA OF THE CHOROID .....       | 270     |
| Morbid Anatomy and Varieties ..... | 272-273 |

|                                                                                        | PAGE    |
|----------------------------------------------------------------------------------------|---------|
| Ophthalmoscopic Picture .....                                                          | 274     |
| Diagnosis .....                                                                        | 274     |
| From Metastatic Carcinoma .....                                                        | 277     |
| GLIOMA OF THE RETINA .....                                                             | 278     |
| Ophthalmoscopic Picture .....                                                          | 279     |
| Morbid Anatomy .....                                                                   | 280     |
| CRYPTOGLIOMA .....                                                                     | 281     |
| PSEUDOGLIOMA .....                                                                     | 282     |
| Ophthalmoscopic Picture .....                                                          | 283     |
| Diagnosis .....                                                                        | 283-286 |
| TUMORS OF THE CILIARY BODY .....                                                       | 287     |
| Sarcoma .....                                                                          | 287     |
| Ophthalmoscopic Picture of Sarcoma of Choroid and Ciliary<br>Body .....                | 287     |
| Less Frequent Tumors of Ciliary Body .....                                             | 288     |
| TUMORS OF THE ORBIT .....                                                              | 289     |
| TUMORS OF THE LACHRYMAL GLAND .....                                                    | 291     |
| TUMORS OF THE OPTIC NERVE .....                                                        | 291     |
| TUMORS OF THE OPTIC NERVE CAUSING OPTIC ATROPHY AND SEC-<br>ONDARY HYPERMETROPIA ..... | 292     |
| DIAGNOSIS OF TUMOR OF THE OPTIC NERVE .....                                            | 293     |

## LIST OF ILLUSTRATIONS.

| FIGURE     |                                                       | OPPOSITE PAGE |
|------------|-------------------------------------------------------|---------------|
| 1          | Ophthalmoscope (obverse) .....                        | 2             |
| 2          | Electric ophthalmoscope .....                         | 2             |
| 3          | Ophthalmoscope (reverse) .....                        | 2             |
| 4          | Ophthalmoscopic examination, indirect method .....    | 5             |
| 5          | Oblique focal illumination .....                      | 5             |
| 6          | Normal fields of vision .....                         | 6             |
| 7          | Skeel's perimeter .....                               | 6             |
| 8          | Duane's tangent plane .....                           | 7             |
| 9          | Right homonymous hemianopsiā .....                    | 7             |
| 10         | Bitemporal hemianopsia .....                          | 7             |
| 11         | Binasal hemianopsia .....                             | 7             |
| 12         | Horizontal hemianopsia .....                          | 8             |
| 13         | Würdemann's transilluminator .....                    | 8             |
| 14         | A sarcoma not demonstrable by transillumination ..... | 8             |
| 15         | Digital tonometry .....                               | 10            |
| 16         | Schiötz tonometer .....                               | 10            |
| 17         | The choroid .....                                     | 12            |
| 18         | The retinal epithelium .....                          | 12            |
| 19         | The layers of the retina .....                        | 13            |
| 20         | The retina at the macula .....                        | 13            |
| 21, 22, 23 | Normal discs with physiologic excavations .....       | 18            |
| 24         | Coloboma of upper lid with coloboma of choroid .....  | 20            |
| 25, 26     | Embryology of Coloboma of the choroid .....           | 22            |
| 27         | Coloboma of choroid .....                             | 25            |
| 28         | Coloboma of choroid .....                             | 25            |
| 29         | Coloboma of optic nerve and sheaths .....             | 25            |
| 30         | Coloboma of optic nerve and sheaths .....             | 31            |
| 31         | Pseudoneuritis .....                                  | 31            |
| 32         | Suppurative choroiditis .....                         | 37            |
| 33         | Choroiditis, stage of exudation .....                 | 37            |
| 34         | Choroiditis, stage of exudation .....                 | 37            |
| 35         | Choroiditis, formation of granulation tissue .....    | 37            |
| 36         | Choroiditis, stage of cicatrization .....             | 37            |
| 37         | Choroiditis, advanced cicatrization .....             | 37            |
| 38         | Choroiditis hyperplastica .....                       | 37            |



| FIGURE |                                                                   | OPPOSITE PAGE |
|--------|-------------------------------------------------------------------|---------------|
| 39     | Choroiditis showing vitreous opacities .....                      | 41            |
| 40     | Choroiditis causing flat detachment .....                         | 41            |
| 41     | Choroiditis, stage of repair .....                                | 44            |
| 42     | Choroiditis, atrophic stage .....                                 | 44            |
| 43     | Choroiditis, final stage of atrophy .....                         | 44            |
| 44     | Exudates in suprachoroidea .....                                  | 44            |
| 45     | Choroiditis, atrophic stage .....                                 | 44            |
| 46     | Choroiditis, atrophic stage .....                                 | 44            |
| 47     | Choroiditis, organized connective tissue of suprachoroidea .....  | 44            |
| 48, 49 | Cicatricial formation in choroid .....                            | 45            |
| 50     | Formation of fibrous tissue in choroid of phthisical eye..        | 45            |
| 51     | Formation of fibrous tissue in the choroid .....                  | 45            |
| 52     | Ossification of the choroid .....                                 | 45            |
| 53     | Tuberculosis of the choroid .....                                 | 51            |
| 54     | Typical miliary tubercle .....                                    | 51            |
| 55     | Complete excavation of disc, non-glaucomatous .....               | 58            |
| 56, 57 | Colloid degeneration of choroid (Drusen) .....                    | 58            |
| 58     | Senile degeneration of retina .....                               | 61            |
| 59     | Rupture of choroid, extending into nerve sheath ....              | 61            |
| 60     | Rupture of choroid, extending into optic nerve ....               | 61            |
| 61     | Sclerotic changes in central artery and vein .....                | 67            |
| 62     | Retinal vein dilated from pressure of sclerosed artery..          | 67            |
| 63     | Obstructive endophlebitis .....                                   | 68            |
| 64     | Nearly complete obstruction of retinal artery .....               | 68            |
| 65     | Concentric narrowing of lumen of central artery .....             | 68            |
| 66     | Dilation of perivascular sheath of sclerosed vessel....           | 68            |
| 67     | Syphilitic round cell infiltration of vascular wall.....          | 68            |
| 68     | Syphilitic cicatricial formation in vascular wall.....            | 68            |
| 69     | Linear hemorrhage in nerve fiber layer of retina.....             | 71            |
| 70 -   | Hemorrhages involving deeper layers of retina .....               | 71            |
| 71, 72 | Hemorrhages involving rods and cones .....                        | 71            |
| 73     | Small hemorrhagic extravasation near macula .....                 | 71            |
| 74     | Angiopathic albuminuric retinitis .....                           | 85            |
| 75     | Angiopathic albuminuric retinitis, cystoid spaces .....           | 85            |
| 76     | Edematous infiltration around macula, albuminuric retinitis ..... | 85            |
| 77     | Retinal discoloration after removal of hemorrhage....             | 85            |

# LIST OF ILLUSTRATIONS.

xvii

| FIGURE   |                                                                              | OPPOSITE PAGE |
|----------|------------------------------------------------------------------------------|---------------|
| 78       | Deep hemorrhage, replaced by cicatricial tissue . . . . .                    | 85            |
| 79       | Granular body in retinal layers, albuminuric retinitis . . .                 | 113           |
| 80       | Sclerotic changes in choroidal vessels, syphilis . . . . .                   | 113           |
| 81       | Nearly complete closure of central artery, endarteritis<br>nodosa . . . . .  | 113           |
| 82, 83   | Closure of central artery, with congestion of choroid .                      | 117           |
| 84       | Closure of central artery, longitudinal section . . . . .                    | 129           |
| 85       | Closure of central artery by active endarteritis . . . . .                   | 129           |
| 86       | Closure of central artery by endarteritis, cell prolifera-<br>tion . . . . . | 130           |
| 87       | Complete closure of both branches of central vein . . . . .                  | 130           |
| 88       | Endarteritic obstruction, converted into connective tissue                   | 130           |
| 89       | Degenerative changes, obscuring nature of obstruction . .                    | 131           |
| 90       | Artefact, obscuring nature of obstruction . . . . .                          | 131           |
| 91       | Endothelial proliferation due to traumatism . . . . .                        | 133           |
| 92       | Degenerative changes following obstruction . . . . .                         | 133           |
| 93       | Retina, two weeks after closure of central artery . . . . .                  | 133           |
| 94       | The retina after closure of central artery . . . . .                         | 136           |
| 95       | Field after closure of inferior temporal artery . . . . .                    | 136           |
| 96       | Collateral branch at termination of thrombosis . . . . .                     | 148           |
| 97       | Canalization of thrombus . . . . .                                           | 148           |
| 98       | Another instance of so-called canalization . . . . .                         | 148           |
| 99       | Central vein closed by phlebitis and thrombosis . . . . .                    | 149           |
| 100      | Sclerosis of central vein . . . . .                                          | 149           |
| 101      | Venous occlusion converted into connective tissue . . . . .                  | 153           |
| 102      | Glaucoma, secondary to venous thrombosis . . . . .                           | 153           |
| 103      | Section of retina destroyed by hemorrhage . . . . .                          | 163           |
| 104      | Closure of branch retinal vein . . . . .                                     | 163           |
| 105      | Typical detachment of the retina . . . . .                                   | 163           |
| 106      | Detachment of retina and choroid . . . . .                                   | 163           |
| 107, 108 | Cystic formation in retinal detachment . . . . .                             | 166           |
| 109      | Detachment of retina (field) . . . . .                                       | 166           |
| 110      | Fibers of the vitreous after hardening . . . . .                             | 171           |
| 111      | Space between vitreous and retina, artefact . . . . .                        | 171           |
| 112      | Shrinkage of vitreous without retinal detachment . . . . .                   | 174           |
| 113      | Spontaneous detachment of the retina . . . . .                               | 174           |
| 114      | Detachment of retina due to contraction—iritidocyclitis . .                  | 174           |
| 115, 116 | Detachment of retina due to thrombus of vena vorticosæ                       | 175           |

| FIGURE        |                                                             | OPPOSITE PAGE |
|---------------|-------------------------------------------------------------|---------------|
| 117           | Congenital detachment of the retina . . . . .               | 175           |
| 118           | Old, degenerated detachment . . . . .                       | 176           |
| 119           | Detachment showing connective tissue degeneration . . . .   | 176           |
| 120           | Retina two weeks after spontaneous detachment . . . . .     | 176           |
| 121           | Ring scotoma, retinitis pigmentosa . . . . .                | 180           |
| 122           | Retinitis pigmentosa (field) . . . . .                      | 180           |
| 123           | Retinitis pigmentosa, flat preparation . . . . .            | 187           |
| 124           | Retinitis pigmentosa, atrophy, except in region of macula   | 187           |
| 125           | Retinitis pigmentosa, degeneration of retinal layers. . . . | 188           |
| 126           | Retinitis pigmentosa, highly atrophic choroid . . . . .     | 188           |
| 127           | Retinitis pigmentosa, simulated by injury . . . . .         | 188           |
| 128           | Retinitis punctata albescens (field) . . . . .              | 193           |
| 129           | Maculocerebral degeneration (field) . . . . .               | 193           |
| 130           | Amaurotic family idiocy; retinal fold, artefact . . . . .   | 207           |
| 131           | Amaurotic family idiocy, optic atrophy . . . . .            | 207           |
| 132           | Comotio retinae, iris pressed against cornea . . . . .      | 207           |
| 133a          | Concentric contraction, wood alcohol poisoning (field) . .  | 209           |
| 133b          | Central tobacco scotoma . . . . .                           | 209           |
| 134, 135, 136 | Retinitis proliferans . . . . .                             | 211           |
| 137, 138      | Retinitis proliferans . . . . .                             | 212           |
| 139, 140, 141 | Choked disc, microscopic sections . . . . .                 | 220           |
| 142, 143      | Choked disc, microscopic sections . . . . .                 | 221           |
| 144           | Choked disc, following injury to cranium . . . . .          | 221           |
| 145           | Optic neuritis in cerebral tuberculosis . . . . .           | 221           |
| 146           | Relation of accessory sinuses to optic nerve . . . . .      | 227           |
| 147           | Ethmoidal contraction of color fields . . . . .             | 227           |
| 148           | Atrophy in post-operative neuritis (field) . . . . .        | 230           |
| 149           | Optic neuritis, post-operative (field) . . . . .            | 230           |
| 150           | Tabetic optic atrophy (field) . . . . .                     | 232           |
| 151           | Minimum contraction, tabetic optic atrophy . . . . .        | 232           |
| 152           | Optic atrophy of tabes, late stage . . . . .                | 234           |
| 153           | Paracentral scotomata in multiple sclerosis . . . . .       | 234           |
| 154           | Oxycephaly, tower skull . . . . .                           | 235           |
| 155           | Round or normally-shaped globe . . . . .                    | 247           |
| 156           | Posterior bulging of globe in myopia . . . . .              | 247           |
| 157           | Myopia with <i>staphyloma verum</i> . . . . .               | 248           |
| 158           | High myopia . . . . .                                       | 248           |
| 159           | High myopia, disappearance of choriocapillaris . . . . .    | 251           |

# LIST OF ILLUSTRATIONS.

xix

| FIGURE   |                                                           | OPPOSITE PAGE |
|----------|-----------------------------------------------------------|---------------|
| 160      | High myopia, thinning of sclera .....                     | 251           |
| 161      | The normal filtration angle .....                         | 251           |
| 162      | Glaucoma, intraocular hemorrhage .....                    | 256           |
| 163      | Glaucoma fulminans .....                                  | 256           |
| 164      | Glaucoma (field) .....                                    | 256           |
| 165, 166 | Glaucoma of long standing .....                           | 258           |
| 167      | Enlargement of ciliary body in advanced life .....        | 258           |
| 168      | Glaucoma, occlusion of the pupil .....                    | 261           |
| 169      | Iris bombé .....                                          | 261           |
| 170      | Total posterior synechia .....                            | 261           |
| 171      | Cyst of pars ciliaris retinae .....                       | 261           |
| 172      | Secondary glaucoma, total anterior synechia .....         | 261           |
| 173      | Secondary glaucoma, lens blocking filtration angle ....   | 261           |
| 174      | Epithelial cyst following corneal wound .....             | 261           |
| 175      | Cystic space in obliteration of anterior chamber .....    | 261           |
| 176      | Glaucoma, epithelium blocking filtration angle .....      | 261           |
| 177      | Glaucoma due to spontaneous dislocation of lens .....     | 261           |
| 178      | Glaucoma due to spontaneous dislocation of lens .....     | 262           |
| 179      | Glaucoma following cataract extraction .....              | 262           |
| 180      | Sarcoma of choroid, drawing iris from cornea .....        | 262           |
| 181      | Glaucoma, due to sarcoma of choroid .....                 | 262           |
| 182      | Glaucoma, due to cyst of iris and ciliary body .....      | 262           |
| 183      | Glaucoma, blocking of angle by cells from cornea and iris | 262           |
| 184      | Old glaucoma, atrophy of iris and ciliary body .....      | 263           |
| 185      | Ciliary staphyloma .....                                  | 263           |
| 186      | Intercalary staphyloma .....                              | 263           |
| 187      | Equatorial staphyloma .....                               | 264           |
| 188      | Early glaucoma, pit-like depressions on the disc .....    | 264           |
| 189, 190 | Early glaucomatous cupping of the disc .....              | 264           |
| 191      | Glaucomatous cupping, later stage .....                   | 264           |
| 192      | Glaucomatous excavation with new connective tissue ....   | 271           |
| 193      | Glaucomatous excavation with new vascularized tissue ..   | 271           |
| 194      | Glaucomatous excavation with fibrous projection .....     | 271           |
| 195      | Buphthalmus .....                                         | 271           |
| 196      | Sarcoma of choroid—stage of extrabulbar extension ...     | 271           |
| 197      | Sarcoma of choroid, projecting into vitreous .....        | 271           |
| 198, 199 | Sarcoma of choroid, projecting into vitreous .....        | 271           |
| 200      | Flat sarcoma of the choroid .....                         | 271           |

## LIST OF ILLUSTRATIONS.

| FIGURE   |                                                                 | OPPOSITE PAGE |
|----------|-----------------------------------------------------------------|---------------|
| 201      | Alveolar sarcoma, endothelioma of choroid .....                 | 271           |
| 202      | Sarcoma of the choroid, spindle-celled .....                    | 272           |
| 203      | Angio-sarcoma of the choroid .....                              | 272           |
| 204      | Alveolar sarcoma of the choroid—endothelioma .....              | 273           |
| 205, 206 | Perithelioma of choroid and ciliary body .....                  | 273           |
| 207      | Sarcoma of choroid, intracellular and isolated pigment .....    | 274           |
| 208      | Sarcoma of choroid, migration of cells along optic nerve .....  | 274           |
| 209      | Small leucosarcoma of the choroid .....                         | 274           |
| 210      | Sarcoma of choroid with equatorial staphyloma .....             | 277           |
| 211, 212 | Metastatic carcinoma of choroid .....                           | 277           |
| 213      | Glioma of the retina, gross specimen .....                      | 280           |
| 214, 215 | Glioma of the retina, microscopic specimens .....               | 280           |
| 216      | Glioma, ganglion and neuroglia cells .....                      | 283           |
| 217, 218 | Metastatic endophthalmitis, umbrella-like detachment..          | 283           |
| 219      | Metastatic endophthalmitis .....                                | 285           |
| 220      | Circumscribed exudate due to injury, simulating tumor..         | 285           |
| 221      | Post-lental membranous exudate .....                            | 285           |
| 222      | Sarcoma of ciliary body, luxating lens .....                    | 288           |
| 223      | Sarcoma of choroid and ciliary body, poorly pigmented .....     | 288           |
| 224      | Conglomerate tubercle of ciliary body, invading lens ....       | 288           |
| 225      | Conglomerate tubercle of ciliary body, perforating sclera ..... | 288           |
| 226      | Metastatic carcinoma of ciliary body .....                      | 288           |
| 227      | Metastatic hydronephroma of ciliary body .....                  | 288           |
| 228      | Cyst of the pars ciliaris retinae .....                         | 288           |
| 229      | Uveal pigment simulating growths in ciliary body .....          | 288           |
| 230, 231 | Telangiectasis of ciliary body .....                            | 289           |
| 232      | Orbital tumor, causing localized hypermetropia .....            | 291           |
| 233      | Tumor of optic nerve .....                                      | 291           |
| 234      | So-called glioma of optic nerve .....                           | 291           |

## LIST OF COLORED PLATES.

|   |                                           | OPPOSITE PAGE |
|---|-------------------------------------------|---------------|
| 1 | Coloboma of the choroid .....             | 20            |
| 2 | Miliary tuberculosis of the choroid ..... | 50            |
| 3 | Rupture of the choroid .....              | 60            |
| 4 | Detachment of the retina .....            | 162           |

## PART II.

### CONTENTS OF PORTFOLIO NO. I.

#### INDEX CARD.

#### DIAGNOSTIC CARDS.

1. Differentiation of Coloboma of the Choroid from Cicatrix of the Choroid; Coloboma of the Optic Nerve from Glaucoma; Congenital Albinism from Acquired Albinism of the Eyeground.
2. Differentiation of Exudative Stage of Disseminated Choroiditis from Albuminuric Retinitis and Diabetic Retinitis.
3. Differentiation of Retinal Opacity of Choroiditis from Retinal Opacity of Arterial Closure; Chorioretinitis Pigmentosa from Retinitis Pigmentosa.
4. Differentiation of Focal Tubercular Choroiditis from Exudative Choroiditis; Albuminuric Retinitis from Chronic Choroiditis.
5. Differentiation of Simple Optic Atrophy from Neuritic Atrophy, Retinitic Atrophy and Glaucoma.
6. Differentiation of Acute Glaucoma from Acute Iridocyclitis and Iritis.
7. Differentiation of Sarcoma of the Choroid from Metastatic Carcinoma of the Choroid.
8. Differentiation of Glioma of the Retina from Pseudoglioma.

#### STEREOGRAMS 1-35 (INCLUSIVE).

1. Use of the Ophthalmoscope. (Direct method.)
2. Normal Fundus Oculi.
3. Retinal Light Reflexes in Negroid Fundus.
4. The Retinal Vessels.
5. Physiologic Excavation of Optic Disc.
6. Coloboma of the Choroid.
7. Coloboma of the Choroid and Optic Nerve.
8. Coloboma of the Macula.
9. Coloboma of the Optic Nerve and Nerve Sheath.

10. Inferior Optic Crescent.
11. Opaque, or Medullated Nerve Fibers.
12. Prepapillary Retinal Artery.
13. Pseudoneuritis and Tortuous Vessels in Hypermetropia.
14. Persistent Canal of Cloquet.
15. Albinism of the Eyeground.
16. Punctate Condition of the Fundus (Gunn's or Crick Dots).
17. Choroiditis Disseminata, exudative stage.
18. Choroiditis Disseminata, exudative stage.
19. Central Syphilitic Choroiditis, exudative stage.
20. Choroiditis Disseminata, cicatricial stage.
21. Central Choroiditis, commencing cicatrization.
22. Chorioretinitis Pigmentosa, Retinitic Optic Atrophy.
23. Tuberculosis of the Choroid.
24. Diffuse Chorioretinitis, atrophic stage.
25. Vascular Degeneration, or Sclerosis of the Choroid.
26. Atrophy of Choroid causing Albinism of the Eyeground.
27. Senile Changes in the Fundus; Colloid Excrescences (Drusen), Macular Degeneration and Arteriosclerosis.
28. Rupture of the Choroid.
29. Diffuse Retinal Arteriosclerosis, primary stage.
30. Advanced Retinal Arteriosclerosis.
31. Arteriosclerosis and Neuritis.
32. Arteriosclerosis with Hemorrhages.
33. Preretinal, or Subhyaloid Hemorrhage.
34. Angiopathic Retinitis, precursory stage.
35. Albuminuric Retinitis, exudative stage.

## CONTENTS OF PORTFOLIO NO. II.

### STEREOGRAMS 36-79 (INCLUSIVE).

36. Albuminuric Retinitis, degenerative stage.
37. Albuminuric Retinitis, atrophic stage.
38. Diabetic Retinitis.
39. Leukemic Hemorrhagic Retinitis.
40. Retinitis Circinata.
41. Closure of the Central Retinal Artery.
42. Closure of the Central Retinal Artery.
43. Closure of Central Retinal Artery with Visible Collateral Circulation and Granular Blood Current.
44. Retinal Atrophy following Closure of Central Artery.
45. Closure of Branch of Central Artery.
46. Closure, or Thrombosis of Central Retinal Vein.
47. Closure of Central Vein with Neuritis, or Hemorrhagic Neuroretinitis.
48. Closure of the Superior Retinal Vein.
49. Post-thrombotic Changes in the Fundus.
50. Incipient, Flat Detachment of the Retina.
51. Well-marked, Recent Detachment of the Retina.
52. Old, Translucent Detachment of the Retina.
53. Retinitis Pigmentosa, Retinitic Optic Atrophy.
54. Retinitis Punctata Albescens.
55. Maculo-cerebral Degeneration, advanced stage.
56. Maculo-cerebral Degeneration, early stage.
57. Amaurotic Family Idiocy.
58. Retina in Congenital Cyanosis, or Cyanosis Retinæ.
59. Traumatic Edema of the Retina, or Commotio Retinæ.
60. Retinitis in Wood Alcohol Toxæmia.
61. Retinitis Proliferans.
62. Retinitis Striata.
63. Papilledema, or Choked Disc in Brain Tumor.
64. Papilledema, or Choked Disc in Brain Tumor.
65. Papilledema in Disease of the Accessory Sinuses.
66. Optic Neuritis in Postoperative Cerebral Sinus Thrombosis.
67. Simple, or Grey Optic Atrophy in Tabes.



68. Simple Optic Atrophy in Multiple Sclerosis.
69. Postneuritic Optic Atrophy.
70. Non-progressive Myopia.
71. Progressive, Pernicious, or Malignant Myopia.
72. Simple Glaucoma, with Total Excavation of Disc.
73. Glaucoma with Advanced Arteriosclerosis, the so-called Hemorrhagic Glaucoma.
74. Leuco-sarcoma of the Choroid.
75. Melano-sarcoma of Choroid with Total Detachment of Retina.
76. Glioma of the Retina.
77. Pseudoglioma, or Metastatic Endophthalmitis.
78. Sarcoma of the Choroid and Ciliary Body.
79. Optic Atrophy and Hypermetropia, secondary to Tumor of the Optic Nerve.

## CHAPTER I.

### EXAMINATION OF THE EYE.

**THE OPHTHALMOSCOPE.** The eye is distinguished from all other organs of the body in that its tissues are transparent. Here can be observed currents of living blood circulating within the vessels, also the secret processes of disease and repair. Within the deep tissues of the eye we may see hemorrhages and exudates appear and disappear, blood vessels pass through the varied stages of degeneration, and neoplasms develop the persistence of which will destroy life. It is unfortunate that these open secrets should be hidden from any practitioner of medicine, particularly, as the presence of many insidious diseases is manifest early in the eye, and frequently the first intimation that nephritis, arteriosclerosis or grave disease of the nervous system is present comes through an ophthalmoscopic examination.

To use the ophthalmoscope successfully is a simple mechanical procedure the attendant difficulties of which have been overrated. The fault with all beginners in ophthalmoscopy is haste. As a matter of fact, no method of physical diagnosis is more readily acquired, provided each simple manœuvre is mastered in proper sequence.

The ophthalmoscope is a small mirror in the center of which is an aperture through which the observer looks. The mirror collects light from a lamp or other source of illumination and reflects it through the pupil, into the eye of the subject (Fig. 1). Electric ophthalmoscopes are now made in which the source of light is a small electric lamp attached in front of the mirror. This arrangement removes the chief difficulty attending ophthalmoscopic examinations, *i. e.*, manipulation of the mirror so as to reflect light accurately through the pupil of the subject. The electric ophthalmoscope illuminates the fundus perfectly in a lighted room and is especially valuable for bedside examinations (Fig. 2).

Where both subject and observer have normal refraction, a perforated mirror is the only instrument required to obtain a view of the fundus. If, however, errors of refraction are present, a correcting glass must be employed. The observer may wear his own glass, but refractive errors in the subject are corrected by a series of lenses attached to the ophthalmoscope in such manner that the requisite lens is easily brought

into the aperture of the mirror (Fig. 3). The beginner in ophthalmoscopy should employ as a subject for examination a young person whose pupils naturally are wide, or someone whose pupils have been dilated by a mydriatic, bearing in mind the danger of using powerful mydriatics on those adults who possess a predisposition to glaucoma.

For descriptive purposes, the several steps of an ophthalmoscopic examination will be presented in the following order: (1) Illumination; (2) Direct examination; (3) Indirect examination.

**ILLUMINATION.** The examination is conducted in a dark room. Both subject and observer are seated. A lamp with a large flame is placed near and slightly behind the subject's head, on the same side as the eye to be examined. For this purpose I prefer the round flame oil or gas burner. The observer, facing the subject at the distance of about one foot, holds the back of the mirror close to his own eye, peeps through the aperture and throws a reflection of the lamplight through the pupil of the subject (Stergm. 1). The observer employs the right eye to examine the right eye of the subject and *vice versa*. The subject should not look directly into the mirror, but just past the head of the observer. As light reflected from the mirror enters the eye of the subject, the pupil presents a brilliant red illumination, due to rays from the fundus reflected into the mirror and, incidentally, through the aperture into the eye of the observer. No fundus details are seen in simple illumination if the eye is normal. Although this procedure is easily performed, it should be thoroughly mastered, inasmuch as manipulation of the mirror in this and other positions is the chief technical factor in ophthalmoscopy. Having acquired sufficient dexterity to hold the fundus reflex with the mirror, the observer rotates a convex lens of 6 D. into the aperture of the ophthalmoscope and approaches the eye of the subject until the red reflex is sharply outlined by the edge of the pupil. To illuminate all parts of the fundus the subject fixes his eye in various positions, *i. e.*, to the right, to the left, upwards, downwards, etc. Simple illumination affords valuable information. Thus, if the media are clear and the fundus normal, the pupil will be uniformly red in all positions of the eye, if, however, opacities exist in the cornea, aqueous, lens or vitreous, they will obstruct the light and appear dark or black. Vitreous opacities usually appear as floating objects, while incipient cataract presents fixed opacities, often arranged like the spokes of a wheel. Gross lesions in the choroid and retina change the color of the fundus reflex. For example, a detached retina gives a pearl-grey reflex, choroidal cicatrices and retinal exudates appear white, while an intraocular tumor



Fig. 1.



Fig. 2.



Fig. 3.

Fig. 1—Ophthalmoscope (obverse). Fig. 2—Electric Ophthalmoscope. Fig. 3—Ophthalmoscope (reverse)



may give no reflex and the pupil becomes black. The strength of the convex lens in the aperture of the ophthalmoscope should now be increased to 12 or even 18 D. and the eye closely approached until details in the iris are in accurate focus. With this glass the finest opacities in the cornea and lens may be seen. Whether a given opacity is in front or behind the iris is determined by the test of parallax displacement. This is performed by noting the relative position which the opacity bears to the margin of the pupil. The observer then slowly moves his head from side to side and the opacity will appear to follow in the same direction if situated behind the iris: on the other hand, it will appear to move in a direction opposite to that of the head if situated in front of the iris. In a like manner the relative positions of any two objects in the fundus may be determined by the parallax test.

The observer next proceeds to examine details in the eyeground. Two principal methods are employed to obtain an ophthalmoscopic view of the fundus, *i. e.*, the *direct*, in which only the ophthalmoscope is employed, and the *indirect*, in which a strong auxiliary lens is interposed between the ophthalmoscope and subject.

USE OF THE OPHTHALMOSCOPE. STEREOGRAM 1. DIRECT METHOD. By the direct method the observer looks into an eye as through an open window and objects in the fundus are seen erect in their natural positions. If both subject and observer have normal refraction, no lens is placed in the aperture of the ophthalmoscope. The lamp remains, as heretofore, at the side of the eye to be examined. The mirror is tilted on its vertical axis toward the lamp so as to reflect light into the pupil. In the electric ophthalmoscope the lamp and mirror are fixed to the instrument in their proper positions. The observer comes very close to the subject, keeping the pupil illuminated, and if the mirror is properly held a distinct view of the eyeground will be obtained. If the subject has small pupils, they should be dilated with a weak mydriatic, such as cocaine, euphthalmine, homatropine, etc. Atropine should not be employed for this purpose, as its prolonged action may precipitate glaucoma in elderly people if a predisposition to this disease exists.

The following rules should be observed in direct examination of the fundus:

- (1) In the primary position, the subject's eye should be directed slightly to the nasal side, which will bring the optic disc in direct line of the observer's vision.

- (2) The observer should keep his head perpendicular with that of

the subject. If held transversely, the forehead will come before the subject's disengaged eye which then, having lost its point of fixation, will move.

(3) The reflection from the ophthalmoscopic mirror casts a small, central shadow, due to the aperture. Guide the light into the subject's eye by keeping this shadow directly on the pupil.

(4) The observer must relax his own accommodation. This is accomplished by looking at the fundus picture as though it were situated at infinite distance. If sought for close at hand, the observer exercises accommodation and the picture will be blurred. Beginners invariably violate this rule.

(5) Hold the mirror close to the subject's eye, as in this way a larger field is obtained.

By the direct method of examination, objects in the fundus are magnified about 15 diameters by the dioptric system of the eye. This is a much greater magnification than can be obtained by the indirect method. The direct method is employed to study vascular changes, location and nature of exudates, measurement of levels and all minutiae in the fundus.

By the direct method of examination, elevations and depressions in the fundus are recognized and accurately measured. For example, a swollen nerve or detachment of the retina has a more hypermetropic refraction than other parts of the fundus and requires the addition of a convex lens to the ophthalmoscope to render it distinct. The strength of the lens necessary to accomplish this measures the height of the elevation, the rule being that 3 dioptries of refraction is equal to a difference in levels of 1mm. The depth of a depression in the fundus, as the floor of a glaucomatous cup, is measured in the same manner, except that in this case a concave lens is added to the combination suitable for other parts of the fundus.

INDIRECT METHOD (Inverted image). In examining the eyeground by the indirect method, the source of light should be slightly above the subject's head. The observer, seated about 18 inches in front of the subject, holds a biconvex lens of 14 to 16 D. at its focal length before the eye to be examined, at the same time, illuminating the eye with the ophthalmoscope (Fig. 4). When properly held, the biconvex lens will intercept the rays of light returning from the subject's eyeground and will form an inverted, aerial image of the fundus between the lens and the ophthalmoscope. It is more difficult to obtain a fundus picture by the indirect than by the direct method, but after a little practice the image







**Fig. 4.**

**Indirect Method of Ophthalmoscopic Examination.**



**Fig. 5.**

**Oblique Focal Illumination.**

suddenly will be perceived and the observer is amazed that it was not seen before. The observer should hold the lens between the forefinger and thumb while the little finger rests on the subject's temple. The plane of the lens should correspond with the plane of the subject's cornea, although if given a slight obliquity certain annoying surface reflexes will disappear. It is desirable to hold the lens in the left hand when examining the right eye, and in the right hand when examining the left eye. The image may be obtained with the open aperture. It is advisable, however, to look through a lens of 4 or 5 D. which will relieve the accommodation of the observer and enable him to come closer to the image. The observer should first find the optic disc, and starting from this point explore the eyeground in all directions. In the indirect image the position of objects is reversed; thus, the upper part of the picture is the lower part of the fundus, the macula appears on the nasal side, etc. By the indirect method a magnification of but three or four diameters is obtained as compared with fifteen diameters by the direct method. Therefore, the indirect method is not the best for examining details; on the other hand, it possesses the following advantages over the direct: (*a*) a much larger field is covered and a more comprehensive view of the eyeground obtained; (*b*) the illumination is much brighter and the fundus can be seen through opacities which would obstruct the view by the direct method; (*c*) it is the only method by which a satisfactory view of the fundus can be obtained in cases of high myopia and astigmatism; (*d*) a fundus picture can be obtained through a narrow pupil; (*e*) patients can easily be examined in the recumbent position.

**FOCAL OR OBLIQUE ILLUMINATION.** Every ophthalmoscopic examination should be preceded by *oblique illumination* of the eye. For this purpose, the source of light should be situated a few feet distant from the eye to be examined. With a convex lens of 15 to 20 D. the light from a lamp or window is concentrated so that the focal point falls obliquely across the cornea (Fig. 5). As the point of light is moved over the surface of the cornea, the most delicate lesions will be revealed. The iris and lens may be examined by this method and even growths situated far forward in the vitreous may be seen. The observer may wear a second convex lens to magnify objects which are illuminated by the oblique method.

In diagnosing diseases of the fundus oculi, the ophthalmoscopic examination should be supplemented by all known methods of detecting disease, particularly modern laboratory tests including those for syphilis and tuberculosis. In this work, however, it is appropriate to refer only

to those methods which lie within the province of the ophthalmologist and neurologist, *i. e.*, examination of the visual field and transillumination of the eye.

The *visual field* is the area in space in which objects are perceived at one time. Accurate vision for detail is limited to a small point in the visual field which corresponds to the fovea centralis (central or direct vision). Outside this small area objects are seen with diminishing distinctness from the center to the periphery (peripheral or indirect vision). Examination of the visual field includes tests for color perception as well as for light. The power to distinguish colors varies in different parts of the retina, thus, colors are recognized throughout the central region, but not at the periphery. Each color has its own normal limitations in the visual field, variations from which are pathologic. The normal field for white and for colors is shown in Fig. 6. Rays of light from an object cross each other before reaching the retina, consequently, the nasal field corresponds to the temporal side of the retina and *vice versa*. The normal eye contains a blind spot (Fig. 6) produced by entrance of the optic nerve. Examination of the visual field can roughly be made as follows: Subject and examiner should be about two feet apart. If, for example, the right eye is under examination, the subject closes the left eye and the examiner closes the right eye. The subject looks steadily into the eye of the examiner while the latter advances his hand, or other object, from periphery to center of field, keeping it equidistant from himself and subject. The subject indicates when the object hand is first perceived, and if the examiner sees it at about the same time, it may be assumed that visual perception exists in that part of the field. The color sense may be examined in the same manner by suitable color cards. These methods answer the purpose for bedside examination or as a primitive test for gross defects, but accurate examination requires the use of a special instrument known as the perimeter (Fig. 7) or a tangent plane chart (Fig. 8). The perimeter consists of a semicircular arc of black metal which revolves upon its center so as to describe the figure of a hollow sphere. A scale of concentric circles is marked on the arc. The subject is so placed that the eye to be examined is in the center of the hollow sphere. The eye is then fixed on the center of the arc while a small white or colored test object is moved along its inner surface from periphery to center and the number of the concentric circle where the test object first comes into view is recorded on a chart. By revolving the arc into all meridians the entire visual field may be plotted. For

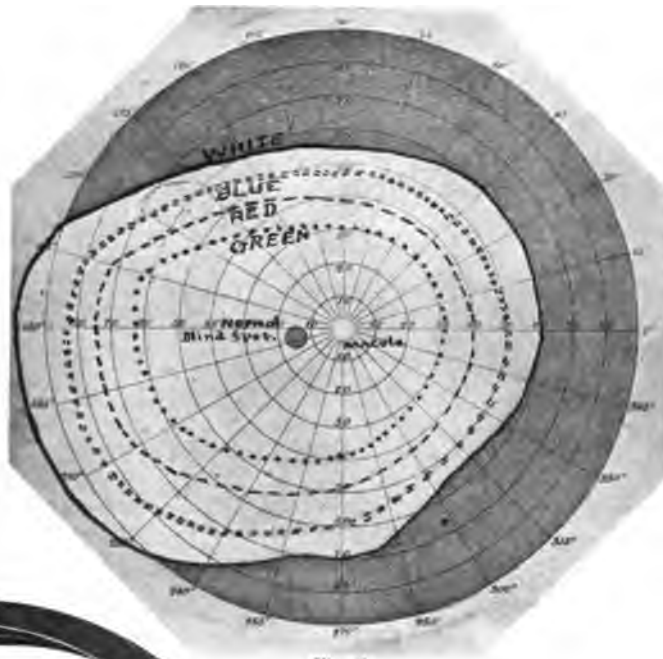


Fig. 6.

Fig. 6—Normal Visual Field for White and Colors. a—Nasal field corresponds to temporal side of retina. b—Temporal field corresponds to nasal side of retina.

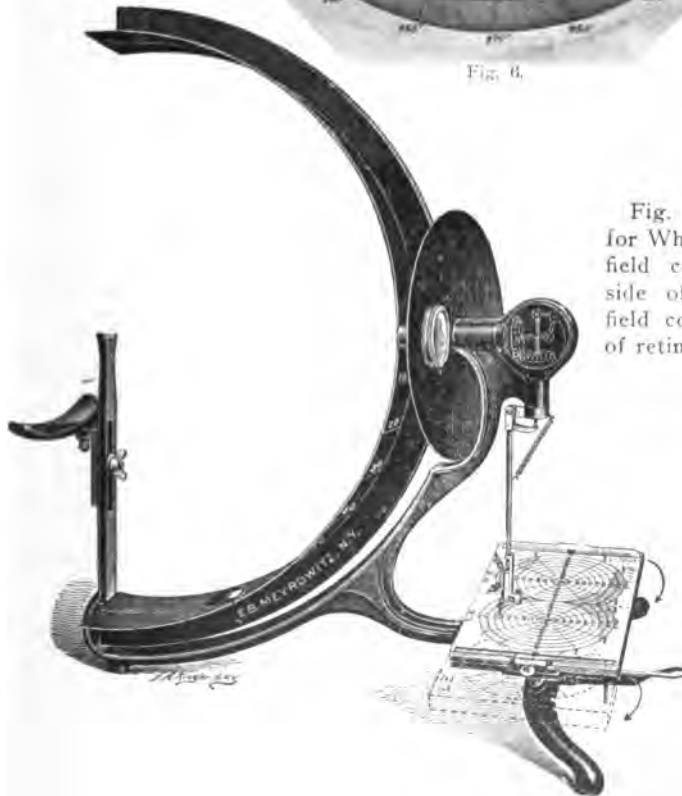


Fig. 7 — Skeel's Perimeter.

Fig. 7.





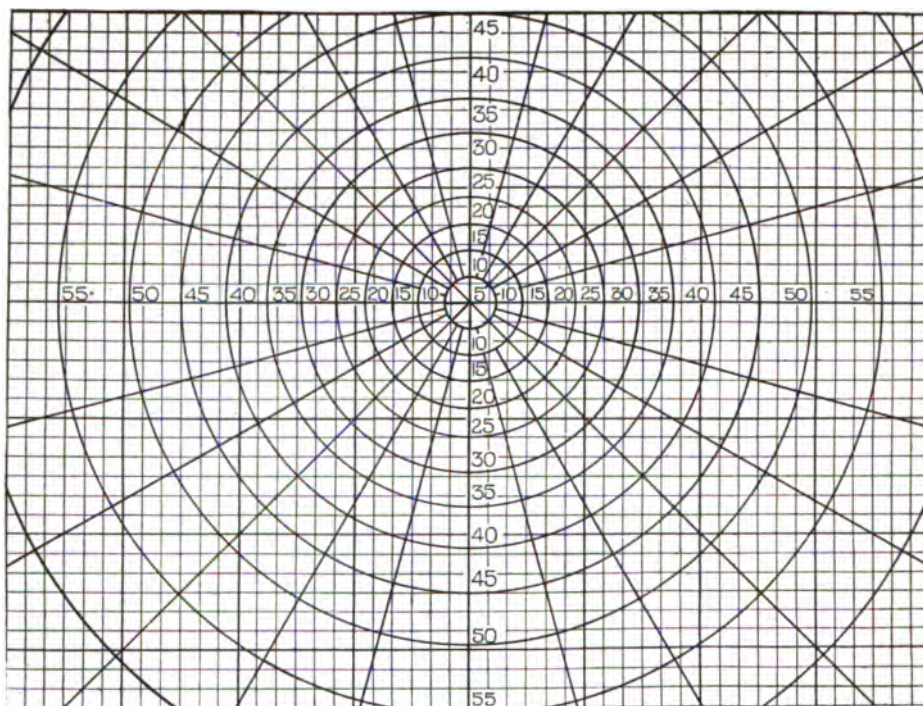


Fig. 8.

Duane's Tangent Plane Chart.

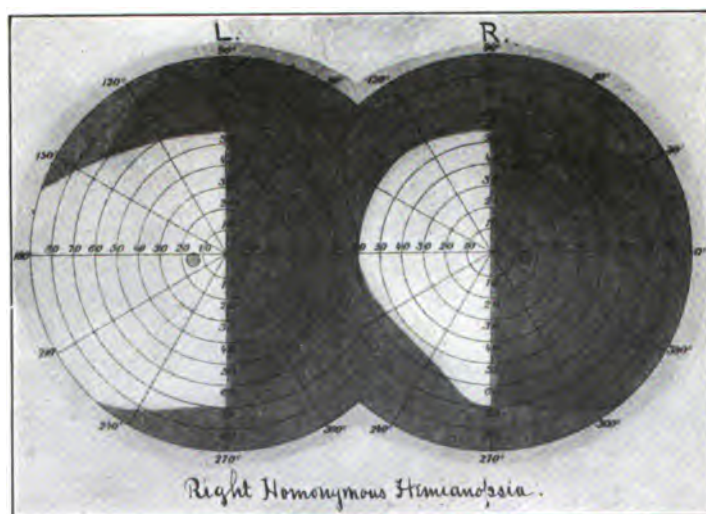


Fig. 9.





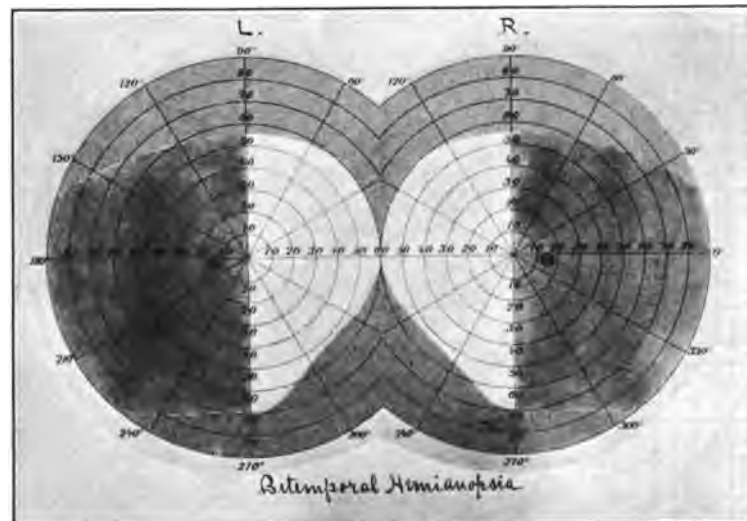


Fig. 10.

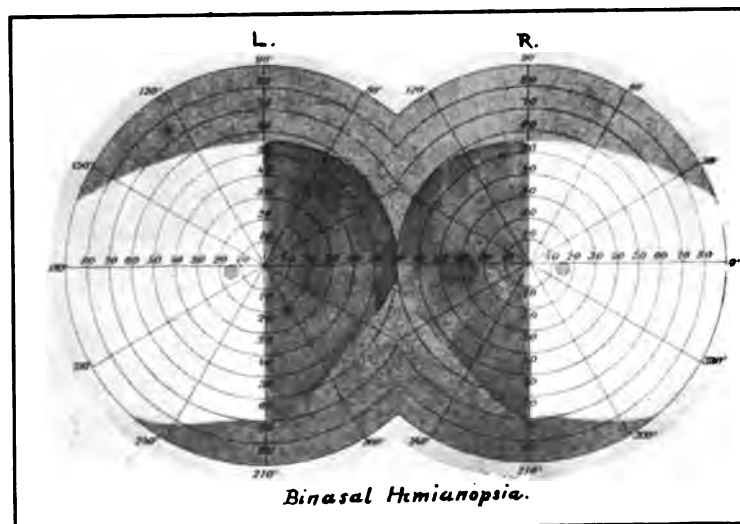


Fig. 11.

mapping out small central or paracentral scotomata a tangent plane (Fig. 8) is better than the perimeter.

A defect in the visual field is known as a *scotoma*. Scotomata are classified according to their situation as central, pericentral, paracentral, equatorial and peripheral, also according to form as, irregular, annular, sector, crescentic, etc. Disappearance of the field from periphery toward the center is called *concentric contraction* (Fig. 150). When one half of the field is destroyed the patient is said to have hemianopia (hemianopsia). The different forms are: (a) Homonymous hemianopia in which vision is lost on the same side, right or left, in each eye (Fig. 9); (b) Bitemporal hemianopia affects both temporal fields (Fig. 10); (c) Binasal hemianopia both nasal fields (Fig. 11); (d) Horizontal or latitudinal hemianopia is the condition in which both upper or lower fields are destroyed (Fig. 12). A scotoma is said to be *positive* when it is perceived by the patient as a black spot or as a cloud in the field of vision. A *negative* scotoma is a blind spot in the field which is not seen by the patient.

**TRANSILLUMINATION.** (Diaphanoscopy.) The interior of the eye may be illuminated, not only through the pupil but also through the sclera, a fact which has been utilized for diagnostic purposes. Special lamps have been devised by Leber, Sachs and Würdemann which, when placed against the sclera, behind the plane of the iris, in a dark room, will light up the interior of a normal eye and render the pupil luminous (Fig. 13). If, however, an opaque object lies between the lamp and the pupil, it will cast a shadow and the pupillary illumination will be darkened. For example, if the transilluminator is pressed against the sclera at a point corresponding to the attachment of a melanosarcoma, the light cannot penetrate the tumor and the pupil remains black. If the transilluminator is then moved to a point not occupied by the sarcoma, the pupil becomes illuminated. Another form of transilluminator, known as the diaphanoscope, is placed in the mouth and pharynx. In this position the light is diffused through the back of the eye, thus revealing opaque objects situated posterior to the equator. Transillumination is the most reliable test we possess for diagnosing sarcoma of the choroid. It may fail, however, under the following conditions: (1) When the aperture in the lamp is not entirely covered by the tumor; (2) A small leucosarcoma may cast little shadow. In such case, if the illumination is reduced a shadow may be demonstrated; (3) In diffuse sarcoma;

(4) When the base of a sarcoma is attached to the posterior pole and the head forms an elongated tumor directed forwards (Fig. 14\*). In transillumination the intensity of the light should be varied, a strong light being required when the globe is filled with blood or contains a dense tumor, while a dim light will be necessary to demonstrate slight variations in the luminosity of the pupil. The following is formulated from Friedenbergs paper on transillumination:

In normal eyes the sclera always is translucent and the transscleral glow illuminates the pupil.

Corneal opacities, however dense, offer no obstruction to the light.

Recent, dense pannus slightly dims the glow.

Uveal pigment in the cornea and elsewhere is an absolute barrier to the light.

No form of cataract offers the slightest obstruction to light in transillumination.

In siderosis of the lens the light is slightly dimmed.

The choroidal pigment is so scattered that it offers no obstruction to transillumination; on the other hand, the closely packed pigment of the ciliary body and iris is an absolute barrier to light and the normal iris is opaque regardless of its color. In atrophy of the iris and in albinos, conditions in which pigment is absent, the light readily passes through the iris stroma. Secondary membranous cataract obstructs light only when it contains a blood clot or uveal pigment. Freshly extravasated blood slightly dims the light but clotted blood entirely obstructs it. Simple detachment of the retina is absolutely transparent. Melanosarcoma of the choroid casts a dense shadow which dims the pupil and darkens the sclera at the site of attachment. Glioma of the retina casts no shadow unless it has undergone calcareous degeneration.† By combined use of the ophthalmoscope and transilluminator the visible area of the fundus is enlarged.

Puncture is sometimes employed to determine the solid or fluid character of retinal elevations. If an aspirating needle is employed it may enter a tumor and withdraw blood and tumor cells. Frequently, however, the needle has failed to encounter a growth when one was present. This

\* From N. Y. Post Graduate Hospital. Service of Dr. A. E. Davis.

† Glioma of the retina casts little or no shadow in transillumination because, (1) glioma is a retinal growth, consequently, a space exists between tumor and sclera through which light enters the eye; (2) glioma contains no pigment.

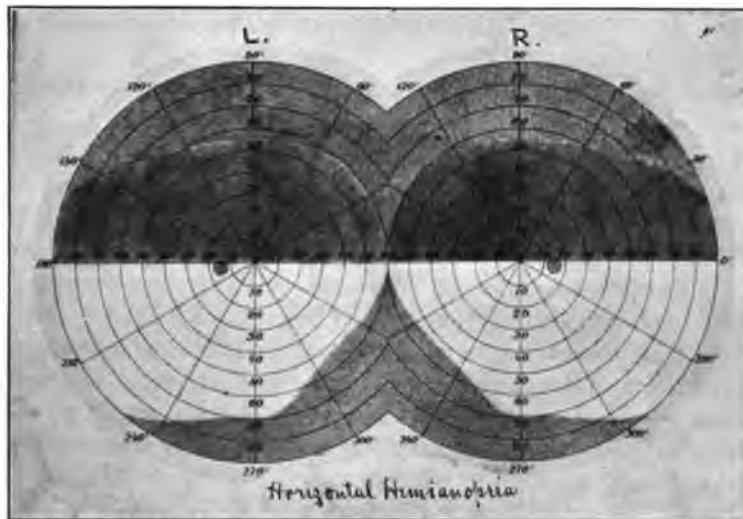


Fig. 12.

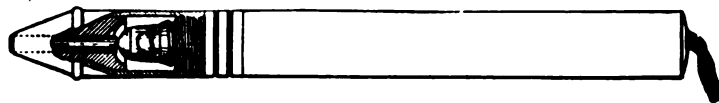


Fig. 13.

Würdemann's Transilluminator.



Fig. 14.

Sarcoma of the Choroid so situated that Transillumination may not establish diagnosis.



method of diagnosis is seldom employed, as there is a well founded belief that the point of puncture affords ready egress to tumor elements.

**INTRAOCULAR PRESSURE.** In every examination of the eye the intraocular pressure should be estimated as a matter of routine.

The eye is a soft capsule filled with fluids under a pressure of from 20 to 30 mm. Hg. (18 to 23 mm. Hg. according to Schiötz). This normal pressure produces the *tension* of the ocular walls and maintains the form of the globe. Any marked variation from this standard is pathologic. Thus, in glaucoma pressure may rise to more than 100 mm. Hg. or it may fall, as in phthisis bulbi, to zero. Intraocular pressure is roughly estimated by the sense of touch, and with accuracy by means of instruments. The tension of the walls varies with intraocular pressure and this may be estimated by palpating the eye with the fingers (digital tonometry). If this simple method is practiced by physicians on healthy eyes they soon acquire the *tactus eruditus* which enables them readily to distinguish any marked variations from the normal tension. The following symbols are employed to record the examiner's impression as to the tension of a particular eye: T is employed to indicate tension; T<sub>n</sub>, = normal tension. The degrees of increased tension (hypertony) are indicated by T + 1, noticeably increased; T + 2, greatly increased; T + 3, Stone-like hardness. In a like manner diminished tension (hypotony) is designated by the degrees of softness as T - 1; T - 2; T - 3.

**DIGITAL TONOMETRY** is performed by palpating the eye. The patient is directed to look slightly downward so as to get the cornea out of the way; then, the tips of the two index fingers are gently placed upon the globe through the closed upper lid and alternately pressed against the eye exactly as in palpating an abscess or tumor for fluctuation (Fig. 15). The eye should not be directed very far below the horizon, otherwise pressure of the extrinsic muscles will increase tension. It is desirable to take tension behind the cornea near the equator, to accomplish which, the back of the examiner's fingers should just touch the margin of the orbit. In palpating the eye the fingers will be steadied if they barely brush each other. Judgment as to variations in the tension of an eye should be controlled by comparison with the fellow eye or with the normal eye of another person. The normal eye dimples under the fingers while in absolute glaucoma it may feel hard as stone.

After instilling a solution of holocaine and directing the patient to look upward, tension can be tested by applying the fingers directly to

the sclera below the cornea. This method is employed when the lids are swollen or the conjunctiva chemotic; under any conditions it is the most accurate method of employing digital tonometry.

INSTRUMENTAL TONOMETRY is performed in two ways:

(1) By the manometer, an instrument which connects the interior of the eye directly with the mercurial column through a canula which perforates the sclera. Obviously, this method, which alone is accurate, cannot be employed clinically.

(2) By the tonometer, an instrument by which intraocular pressure is estimated from the resistance which the walls offer to external indentation. Among the various instruments designed for this purpose only that of H. Schiötz has come into general use (Fig. 16). This instrument is applied to the cornea after the latter has been anesthetized with a 2 % solution of holocaine which does not dilate the pupil. It is a reliable index of intraocular pressure and should be employed in all doubtful cases.



Fig. 15.  
Digital Tonometry.

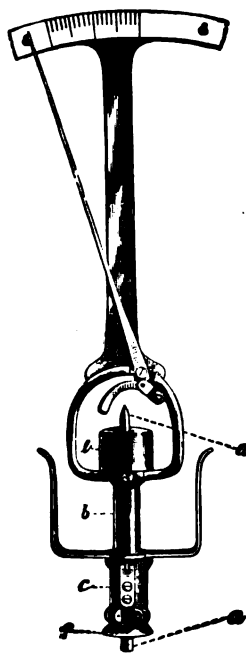


Fig 16.  
Schiötz Tonometer.





## CHAPTER II.

### THE NORMAL FUNDUS.

**STEREOGRAM 2. BLOND FUNDUS.** Right eyeground of a healthy, light complexioned, Swedish girl. The fundus color is a bright yellowish red, indicating that pigment is scanty in both retinal epithelium and choroid. In the macular region and around the nerve the fundus darkens. The macula exhibits a visible foveal reflex. The choroidal vessels are indistinctly outlined but may be recognized as red, anastomosing bands darker than the intervascular spaces. No vessels are seen at the macula. The optic disc is nearly circular in form and of normal color. Both choroidal and scleral ring are well defined. The retinal vessels are distributed in the most conventional manner. A small cilioretinal artery emerges from the temporal side of disc and is directed toward the macula.

The fundus oculi comprises that portion of the eyeground which can be seen with the ophthalmoscope. In the normal eye this includes a little more than the posterior hemisphere. The ophthalmoscopic picture of the fundus is created by light reflected from the coats of the eyeball. The chief factor in its production is the choroid and, to a less extent, the sclera and retina.

**SCLERA.** The sclera is composed of white fibrous connective tissue which, though obscured by the red choroid, furnishes a brilliant white background which lightens the color of the entire fundus.

**SUPRACHOROIDEA.** The sclera is connected with the choroid by the suprachoroidea or lamina fusca, a loose membranous tissue arranged to act as lymph spaces. In disease these spaces may be enormously distended by exudates or blood. The outer layer of the lamina fusca contains considerable pigment, some of which usually remains attached to the sclera after the choroid has been destroyed by disease. The suprachoroidea exerts little influence upon the ophthalmoscopic picture except when distended by pathologic effusions (Fig. 44).

**CHOROID.** The brilliant red coloring of the caucasian eyeground is produced by the choroid. The choroid is the vascular coat of the eye. The blood vessels are arranged in a manner peculiar to this membrane

in three superimposed layers (Fig. 17). The *external layer* consists of the larger vessels, mostly veins, which anastomose freely. The vessels are united by a delicate stroma containing pigmented cells, called chromatophores. The *middle layer* is made up of medium sized vessels. The stroma contains but few pigmented cells. The *inner layer*, termed the *choriocapillaris*, consists entirely of closely arranged, wide capillaries. Pigmented cells never appear in the choriocapillaris under normal conditions. On its inner surface, the choroid is separated from the pigmented epithelial layer of the retina by a homogeneous, elastic membrane; the *lamina vitrea*. On the inner surface of the lamina vitrea the retinal epithelium forms a close mosaic of pigmented, polygonal cells (Fig. 18). This so-called "retinal" epithelium develops from the same blastodermic layer as the retina, but anatomically and pathologically it is a part of the choroid. The color and brilliancy of the ophthalmoscopic image is modified by the relative amount of pigment contained in the retinal epithelium and chromatophores of the choroid. Usually, the amount of pigment in the epithelial layer corresponds to that in the choroid, but disproportionate pigmentation of the two membranes is not uncommon. As a rule, the fundus of a blond individual is light in color and of a brunette dark, but exceptions are frequent. The following normal variations in fundus pigmentation produce the more familiar types of eye-grounds.

(1) Excessive pigmentation of both choroidal cells and retinal epithelium, as in negroes, produces a poorly illuminated fundus, slate colored at the posterior pole and chocolate color at the periphery (Strgm. 3).

(2) A moderate amount of pigment in the retinal epithelium is sufficient to hide the choroidal vessels and still permit the passage of a brilliant reflex from the choroid. In such eyes the fundus has a uniform, rich red color (Strgms. 16-29-33-58).

(3) If the retinal epithelium is well pigmented and the choroid poor in pigment, the epithelium becomes visible and the fundus appears darkly stippled or "peppered" (Strgms. 39-67).

(4) If the retinal epithelium is poor in pigment, the choroid can be seen. In such case, if pigment is abundant in the intervascular spaces of the choroid, a tessellated or tigered fundus is produced and the choroidal vessels are of lighter color than the background (Strgm. 55).

(5) If both epithelium and choroid are poor in pigment, the choroidal vessels are darker than the background and the fundus appears reticulated (Strgms. 2-19-56).



Fig. 17.

The Choroid.

(a) external layer, (b) middle layer, (c) internal layer.

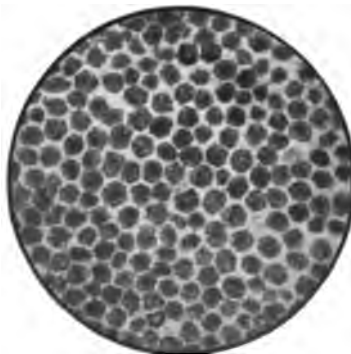


Fig. 18.

The Retinal Epithelium.







Fig. 19.

The Layers of the Retina.

(a) retina, (b) choroid, (c) sclera.

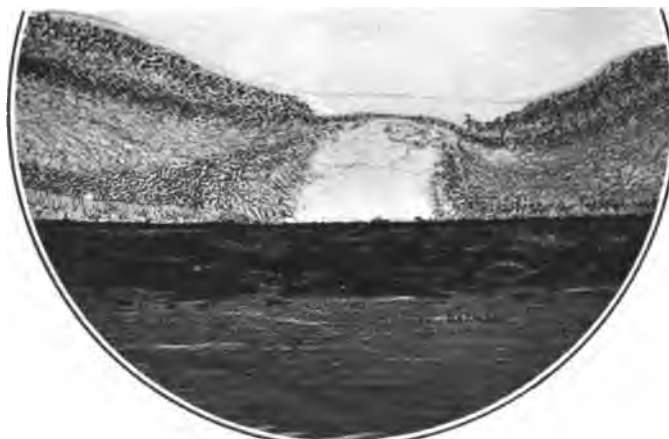


Fig. 20.

The Retina at the Macula.

Note thickness of layer of ganglion nerve cells, a and a'.

(6) In albinos and cases of fundus depigmentation, the fundus contains no pigment and the choroidal vessels are distinctly seen sharply defined against a yellowish-white sclera (Strgms. 15-26).

Vessels of the choroid are easily distinguished from those of the retina by the following peculiarities:

(a) They increase in distinctness and do not narrow toward the periphery.

(b) They are larger and more numerous than retinal vessels.

(c) Their courses are erratic. Groups of vessels frequently run parallel.

(d) They anastomose with each other.

(e) Choroidal vessels contain no central light streak and arteries are indistinguishable from veins.

**RETINA.** The retina is the inner or, as viewed with the ophthalmoscope, most superficial coat of the eye. It is a transparent membrane contributing little toward the normal coloring of the eyeground. The retina simply lies upon the pigmented epithelium, its only attachments being around the optic nerve, and at its anterior border where it joins the choroid by a jagged line called the *ora serrata*. Ten layers are recognized in the retina, of which the five outer are termed neuroepithelial layers and the five inner cerebral layers (Fig. 19). The nerve elements of the retina are supported by a connective tissue framework of ectodermic origin. The principal members of this framework are the fibers of Müller which extend through the retina from the internal limiting membrane to the layer of rods and cones, where they expand and form the external limiting membrane. The *macula lutea*, the center of the fundus, is situated about two disc diameters to the temporal side of and slightly below the optic nerve. This most highly specialized portion of the visual organ is the region of acute vision. The macula is a round or horizontally oval depression, about 2 mm. in diameter, in the surface of the retina. The macula is formed by the following modifications in the structure of the retina. As the retina approaches the border of the macula the layer of ganglion cells increases in thickness from a single-celled layer to a layer eight or more cells deep (Fig. 20). Within the macula, however, all layers of the retina fuse and decrease in thickness until at the fovea centralis, or center of the macula, the membrane consists of little more than a layer of cones and nuclei.

For descriptive purposes the eyeball is compared to the earth: thus, it is said to possess an anterior and a posterior pole between which is



situated the equator. It is further divided by meridians, as the horizontal and the vertical meridians. In conformity with this method of orientation the fundus is separated into ill-defined zones, *i. e.*, a posterior polar zone, a posterior median zone and an equatorial zone, etc.

STEREOGRAM 3. NEGROID FUNDUS. RETINAL LIGHT REFLEXES. Left eyeground of a healthy mulatto boy, 9 years of age. The color of the fundus is characteristic of the dark-skinned races and is due to an excess of pigment in the retinal epithelium and choroid which obscures the choroidal reflex. The optic disc and retinal vessels are normal. This fundus is remarkable for the number and brilliancy of the light reflexes which play over its surface and along the vessels. Bright, silvery rays radiate in a circle from the optic disc corresponding in their courses to those of the nerve fibers. The edge of the foveal cup is marked by a semicircle of light which changes its position with every movement of the mirror.

Although the retina is a transparent membrane, usually invisible, it will reflect light, especially if surface inequalities exist. Visible light reflections from the retina are common in children whose eye media, as a rule, are perfectly transparent. They are also well marked in dark eyegrounds. Retinal reflexes are best seen with a concave mirror and feeble illumination. Usually, they disappear in adult life. Wide variations exist in their form and intensity. The most common examples consist of fine silvery lines around the disc or flashes of pale light which play over and along the vessels. They are a normal phenomenon and their presence usually indicates a healthy fundus. As the ophthalmoscopic mirror is moved, retinal reflexes alter their shape and position and in this way are differentiated from pathologic changes which are fixed. The macula being a concave reflecting surface is rich in reflexes and correspondingly difficult to examine, because, in a full front view, both macular and corneal reflexes fall directly into the observer's line of vision. Examination of the macula is accomplished by the direct method. The subject is told to look at the light in the mirror. At first the observer sees little or nothing, but by bringing a concave lens of 0.5D. into the aperture of the ophthalmoscope, and gazing steadily for a few moments, the fovea will gradually appear. The brightest macular reflex is that which circles around the walls of the foveal depression. Viewed from either side, this reflex appears as a small crescent the concavity of which is always towards the observer; but viewed directly from in front, it appears as a small ring with a diameter equal to that of the largest vessel on

the disc. The macula is darker than other parts of the fundus and the fovea is the darkest part of the macula. If the fundus is deeply pigmented, the macula is uniformly dark, but if the retinal epithelium contains little pigment, the macula as well as other parts of the fundus may be stippled. It is important to differentiate the normal stippling of a light macula from the patchy discoloration of central, syphilitic choroiditis.

**STEREOGRAM 4. THE RETINAL VESSELS.** Left eyeground of a youth in whom the retinal vessels are normal in appearance, and their method of distribution conforms to the usual scheme. This scheme is followed, in a general way, by the majority of eyes but varies widely in individual cases. The disc is normal and contains a large physiologic excavation. Retinal arteries are distinguished from veins as follows. The arteries are narrower than the veins and pursue a straighter course. The surface of the retinal vessels reflects a central streak of light the same as any polished cylinder. On the arteries the central light streak is broad, bright and reddish in color. On the veins it is narrow, dull and white. There has been much inconclusive discussion as to whether the central light streak is due to reflection of light from the walls of the vessel, from the column of blood or from the choroid. The distinctions described apply only to large and medium sized vessels. The character of a small vessel must be determined by tracing it back to its source. Retinal vessels branch dichotomously and diminish in size from the disc to the periphery. In this eye, as the central artery emerges from the disc it divides into the ascending and descending papillary arteries. Frequently, this primary division occurs within the nerve. The ascending artery bifurcates to form the superior temporal and superior nasal arteries. In a similar manner the descending artery splits into an inferior temporal and inferior nasal branch. The veins correspond in name and plan of distribution to the arteries. In the majority of cases the artery emerges from the disc on the nasal side of the vein. Several small vessels emerge from the edge of the disc. The temporal arteries, which are slightly larger than the nasal, sweep in wide curves around the macula to which they send their principal branches. Although devoid of visible vessels, the macular region is richly supplied with capillaries except at the fovea centralis which is absolutely nonvascular. In fact, retinal vessels in this location would be superfluous inasmuch as the fovea consists only of cones and these are nourished exclusively by the choroid. Springing from the temporal side of the disc is a small artery distributed to the region between the nerve and macula. This is called a cilioretinal artery because it is

supposed to arise from the circle of Zinn or from other vessels connected with the choroid. A true cilioretinal artery is recognized by the manner in which it hooks over the edge of the disc, the stem being directed outward toward the choroid. Exceptionally these arteries appear on the nasal side of the nerve (Strgm. 12). The existence of cilioretinal veins is doubtful. If they exist they must arise directly from the choroid as the posterior ciliary system is strictly arterial. The majority of vessels which emerge from the margin of the disc arise from the central vessels far back in the nerve. In case the central artery is occluded, a cilioretinal artery may permanently preserve a small, central field of vision. A rare vascular anomaly, not present in this eye, is an optico-ciliary vessel which arises from a central retinal vessel and passes to the edge of the disc into which it disappears to join the choroid. The retinal vessels are distributed only to the inner layers of the retina. The external layer of the retina which consists of the rods and cones is nourished by the choroid. The trunk vessels are the central artery and central vein of the retina which enter the eye *via* the optic nerve. The central artery arises from the ophthalmic and enters the optic nerve, in company with the vein, about 10-12 mm. behind the eyeball. The central vein of the retina usually empties into the superior ophthalmic. The ophthalmic vein discharges into the cavernous sinus but also communicates with the facial vein. This communication between the ophthalmic and facial veins is of great importance inasmuch as it provides a free outlet for the retinal blood should the cavernous sinus become clogged. In the head of the optic nerve the central retinal artery anastomoses with vessels from the choroid as follows. Just before the short ciliary arteries enter the choroid they give off small branches which unite and form a ring in the sclera around the optic nerve. This arterial ring is known as the *circle of Zinn*. Branches from Zinn's circle enter the head of the nerve and anastomose with twigs from the central artery of the retina. These anastomoses are little more than capillaries; nevertheless, they provide a feeble collateral circulation for the retina in closure of the central artery. Vessels from the central artery supply the nerve and furnish a rich capillary plexus which gives color to the optic disc. After emerging from the disc, the retinal vessels form a strictly terminal system, *i. e.*, they do not anastomose with other vessels or with each other. The retinal vessels are provided with perivascular sheaths which are the lymph channels of the retina.

**PULSE PHENOMENA.** Pulsation of a vein upon the optic disc is a physiologic phenomenon and is present in, approximately, 75 per cent of

normal eyes. Donders explained its occurrence as follows. The cardiac systole drives the blood wave into the eye, producing momentary increase in intraocular pressure, and this rhythmic increase of pressure may be sufficient to empty the veins on the disc. Possibly, venous pulsation may be assisted by pulsatory pressure variations in the cavernous sinus. In accordance with the above theories, the physiologic venous pulse is diastolic, *i. e.*, the veins fill during cardiac diastole and empty as the pulse wave reaches the eye. Another explanation of the venous pulse is that either in the nerve or on the disc, the artery presses against the vein and arterial pulsation is transmitted by contact. This would explain why pulsation is present in some cases and absent in others, and why it frequently is limited to a single vein. Certainly, pulsation always occurs in a vein which is visibly in contact with an artery. Venous pulsation can be produced in normal eyes by lightly compressing the eye and, if the pressure is increased, pulsation appears in the artery and the vein collapses. Pulsation of the retinal veins may be of pathologic import. A positive or systolic venous pulse may occur in tricuspid regurgitation. In aortic regurgitation both veins and arteries pulsate. Pulsation in the retinal arteries always is pathologic.

The systemic blood pressure is just enough higher than intraocular pressure to permit a smooth flow of blood through the retina without visible pulsation of the arteries. If, however, either intraocular pressure rises above or systemic blood pressure falls below its relative standards, the inflow of blood is hindered and the artery will pulsate. Two forms of arterial pulsation are recognized, (1) the pressure pulse which appears only in the arteries on the disc. It occurs in the heightened eye pressure of glaucoma and in the lowered blood pressure of syncope, ischemia, etc. (2) The locomotion pulse, which is manifested throughout the retina by a distinct motion of the arteries seen wherever they make a quick bend. Locomotion pulse occurs in aortic regurgitation, where it is accompanied by a pulsation of the veins and by an alternate flushing and paling of the disc. It may occur also in aneurism, Basedow's disease and in the earliest stage of diffuse arteriosclerosis when the arteries are relaxed (Sternberg, 29). Pulsatory phenomena may be simulated by slight motions on the part of observer or subject, and the diagnosis of abnormal pulsation should never be made unless it is distinct and unmistakable.

STEREOGRAM 5. THE OPTIC NERVE. OPTIC DISC WITH PHYSIOLOGIC EXCAVATION. Right eyeground of a boy 12 years of age. The case was selected for illustration as a good example of the normal disc.

The outline of the optic disc may be either circular or oval. The long diameter usually is vertical or slightly tilted; exceptionally, it lies transversely. The general color of the disc is pink, the temporal side being lighter in color than the nasal; the outline is somewhat softened by the overlying nerve fibers, but is well defined in normal eyes. The majority of nerve fibers pass to the nasal side of the retina; hence, this side of the nerve is less distinct in outline than the temporal; for the same reason the disc is darker and redder on the nasal side. The choroid and sclera usually contain an excess of pigment massed around the edge of the disc, forming a more or less complete black circle called the *choroidal ring*. Immediately within the choroidal ring is a second ring of white connective tissue which is said to represent the distal end of the pial nerve sheath. It is generally called the *scleral ring* from its supposed connection with the sclera. In the stereogram the connective tissue ring appears well developed, but often it is incomplete or absent. The lightest colored part of the nerve-head is centrally located and marks the site of a deep depression, the vascular funnel, a canal which transmits the retinal vessels. Usually, the vessels appear to sink and fade away into the semitransparent tissue which fills the depression. Not infrequently, however, this vascular funnel is very wide and, as in this nerve, occupies a large part of the disc forming a cavity known as a *physiologic excavation* (Figs. 21-22-23). The formation of a physiologic excavation depends upon a sclero-choroidal canal which is too large for the nerve; consequently, the nerve fibers are distributed around the wall of the canal leaving a central space. Physiologic excavations vary in shape and size. The most frequent form is a circular, central depression (Stergm. 40). An oblique, funnel-shaped excavation, opening towards the macular side, as shown in the stereogram, also is common. Deep excavations extend to the lamina cribrosa the perforations of which appear as grey dots on a white surface. The main retinal vessels usually climb up the nasal side of a physiologic excavation. The macular vessels, however, may ascend along the temporal side. When the center of a disc occupies a lower level than the margin the fact may be demonstrated by the test of parallax displacement as follows: with the direct method of examination, if the observer moves his head from side to side, the bottom of the cup will follow while the edge of the cup will move in the opposite direction.

**ANATOMY.** The optic nerve is composed chiefly of axis cylinders or neurons from the ganglion cells of the retina. These neurons, which constitute the inner or nerve fiber layer of the retina, converge to the



Fig. 22.



Fig. 23.

Figs. 21, 22, 23—Normal Discs with Physiologic Excavations.



scleral foramen where they form the optic nerve and pass to the brain. The function of the optic nerve is to convey to the higher cerebral centers the impressions gathered by the retina. The nerve also contains fibers which convey impulses from the brain to the eye. The optic nerve may be divided into four parts: (1) The *intracranial*, which is about 1 cm. in length and extends from the chiasm to the inner opening of the optic canal. This portion of the nerve is subject to pressure from tumors and to constriction from organized inflammatory exudates; (2) The *intra-canallicular* division, which lies within the optic canal. This canal consists of a bony portion 4 mm. in length and a fibrous portion which extends into the cranial cavity for a distance of 2 mm. Only the optic nerve and ophthalmic artery pass through the optic canal, and it is said that sclerosis of the artery may cause pressure atrophy of the nerve. The nerve is injured in the canal also by periostitis, thickening of the bone and, especially, by fracture of the orbit which tends to pass through the optic foramen. (3) The *orbital* division, about 30 mm. in length, includes that portion of the nerve which extends from the optic foramen to the eyeball. This section of the nerve is very lax and pursues an S-shaped course which permits free rotation of the eye without rendering the nerve taut. (4) The *intraocular* division of the nerve comprises that portion of the nerve which pierces the sclera and choroid. It measures about 1.25 mm. in length. The so-called scleral foramen, through which the nerve passes into the eye, consists of numerous small holes in the inner layers of the sclera, forming a sieve-like partition known as the *lamina cribrosa*. The external fibers of the sclera do not enter into the formation of the lamina but are reflected backward and become part of the dural sheath of the nerve. Inasmuch as the lamina cribrosa is composed of unyielding fibrous tissue, it is instrumental in causing congestion and edema of the nerve-head. That portion of the optic nerve situated in front of the lamina cribrosa is ophthalmoscopically visible and constitutes the *optic disc* or *papilla*. The name "papilla" is a survival of early ophthalmoscopic days, when the head of the nerve was erroneously supposed to project into the eye. In normal eyes the optic disc is level with—or below the surrounding fundus. In the optic canal and orbit the nerve is enclosed by extensions of the cerebral membranes, *i. e.*, the dura, arachnoid and pia. The central artery and vein of the retina enter the nerve about 10 mm. behind the globe: therefore, compression or injury of the nerve posterior to this point does not affect retinal circulation.



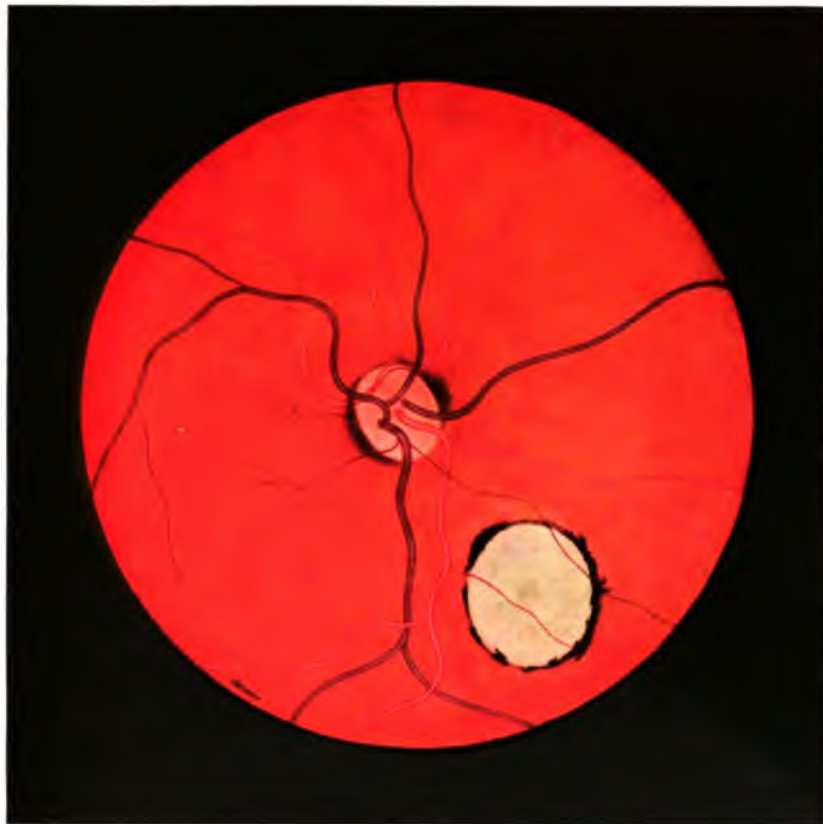
### CHAPTER III.

#### CONGENITAL ANOMALIES.

**STEREOGRAM 6. COLOBOMA OF THE CHOROID.** Right eyeground of boy 12 years of age. Left eye is normal. On his right side the patient has rudimentary auricle, paresis of the seventh nerve, coloboma of the iris, lens and choroid. Contrary to the rule, the coloboma of the choroid is nearly circular in outline. It presents an extensive, bluish-white surface extending from the center of the disc downward and outward. The outward deviation appears to be caused by rotation of the eyeball. The lower half of the optic disc is included in the imperfection. The adjacent fundus is strongly pigmented. The coloboma forms a craterform depression in the fundus about 2 mm. (6.D.) below the normal level. On the floor of the depression are two small, oval ectasiæ. The neighboring fundus, including the optic nerve, appears to slope into the pit. The retinal vessels make wide detours and skirt along the margin of the coloboma as though they had been pushed aside. The defect is covered by retinal tissue, as shown by a medium-sized retinal vessel which crosses from above downward and outward. The branching vessel which enters the coloboma through the floor above, probably is a ciliary artery, while the vascular loops which push in along the edge are from the neighboring choroid. There appears to be dim perception of light throughout the colobomatous area, indicating the presence of retinal elements.

Coloboma of the choroid is a congenital gap in the membrane due to arrested development. The sclera is exposed in the defect and presents a brilliant white or bluish-white surface on which spots of pigment are not uncommon. The typical coloboma of the choroid is situated near the median line below the optic nerve. The smallest coloboma I ever saw was about twice the diameter of the optic disc (colored plate 1), and the largest covered nearly half the fundus. As a rule, a bridge of choroidal tissue separates the defect from the disc, but not infrequently the nerve also is involved. Coloboma of the choroid usually occurs in connection with coloboma of the iris or other malformation (Fig 24). Coloboma of the lens is often associated but usually is overlooked, as it may consist of only a slight flattening at the lower edge. Frequently, the cornea is vertically

**COLORED PLATE I**



**Coloboma of the Choroid**





Fig. 24.

Coloboma of the Upper Lid which was associated with Coloboma of the Choroid.



oval or pointed below. Researches have shown that coloboma of the choroid is eminently hereditary. The typical outline of the imperfection is a vertical oval or an obtusely angled triangle with the apex toward the disc. Rarely it is round or the long diameter is directed transversely. In some fetal eyes both choroid and sclera are so defective that the intraocular tissues grow outward and form a large sac below the eye, while the eyeball remains very small (*microphthalmus with sac*). Eyes with coloboma of the choroid usually are small. The refraction, however, is myopic owing to the ectasia of the sclera. As a rule, some perception of light exists in the colobomatous region.

The diagnosis of typical coloboma of the choroid presents no difficulties, but atypical cases cannot always be differentiated from old cicatrices.

#### COLOBOMA OF THE CHOROID.

Congenital malformation.  
Is below the level of the fundus.  
Usually, a single defect.  
Situated below nerve near median line.  
Always much larger than the optic disc.  
Outline symmetrical. Usually, vertically oval.  
Frequently associated with coloboma of iris nerve or other ocular malformation.  
Usually represented by relative scotoma in visual field.  
Seldom affects both eyes.

#### CICATRIX OF THE CHOROID.

Results from disease or trauma.  
Is level with fundus.  
Usually, more than one lesion.  
Situated anywhere in the fundus.  
May be of any size.  
Usually circular or irregular in outline.  
Not associated with congenital defects.  
Represented by absolute scotoma.  
Frequently present in both eyes.

In typical rupture of the choroid a crescentic area of sclera is exposed, concentric with the optic disc. Rupture is further differentiated from coloboma by a history of injury.

STEREOGRAM 7. COLOBOMA OF CHOROID AND OPTIC NERVE.\*  
Russian girl 11 years of age. Right eye normal. Left eye presents an enormous coloboma of the choroid which includes the lower third of the

\* From the service of Dr. J. Scott Wood, Brooklyn Eye and Ear Hospital.

fundus, the optic nerve and the macula. Its posterior extremity extends beyond the optic nerve and its anterior border beyond the limits of the ophthalmoscopic field. The edge of the coloboma is sharp cut as though the choroid had been punched out with a die. It is framed by a border of dense pigment, and pigment anomalies are seen in the surrounding fundus. The floor of the coloboma is about 1 mm. (3.D.) below the level of the fundus. Its surface is broken by several pits and channels. The entire coloboma is covered with loose, web-like tissue, evidently imperfectly formed retina. Posteriorly, this tissue is thick and contains retinal vessels; anteriorly, it is tenuous and transparent. Filmy membranes and dense tangles of tissue containing pigment extend out from the margin, and web-like bands stretch across the gap. The optic disc is not apparent, but its site is indicated by the retinal vessels which converge to a point in the dense tissue of the coloboma. These vessels can be traced from their origin, across the pigmented border, out into the retina. In the lower part of the coloboma, the large vessel which emerges from the pigmented border and extends across the pit is known, from its manner of branching, to be from the choroid.

**PATHOGENESIS.** Various theories have been advanced to explain coloboma of the choroid, none of which have been established. The situation of the defect, along the median line below the disc, corresponds to the fetal cleft in the secondary optic vesicle. Hence, it was assumed that coloboma of the choroid resulted whenever this cleft failed to unite. Unfortunately for this simple theory, the secondary optic vesicle represents the retina and the choroid does not develop from this layer (Fig. 25). Furthermore, the secondary optic vesicle is situated internal to the choroid and has been found perfectly united in many cases of coloboma of the choroid. The choroid develops from the blastoderm, forms a vascular coat external to the retina, and possesses no fetal cleft (Fig. 26). Another argument against the theory that coloboma is due to nonclosure of the fetal cleft is the occasional occurrence of coloboma on one side of, or even above, the nerve. v. Hippel calls attention to the influence of the vitreous upon the formation of coloboma. If the vitreous is scanty the retina does not get spread out but lies in folds some of which may fall into the fetal cleft. This accident or the persistence of mesoblastic tissue in the cleft, is sufficient to prevent development of the choroid at this point. Treacher Collins and W. Lang attribute all cases of choroidal coloboma to abnormal adhesions between the mesoblastic choroid and epiplastic retina. Prenatal inflammation has been widely ac-



**Fig. 25.**

**Secondary Optic Vesicle. A, exterior layer; P, interior layer; M, mesoderm; L. lens.**



**Fig. 26.**

**Development of the Choroid from the Blastoderm.**





cepted as a cause for coloboma, but microscopic examinations have failed to discover any evidence of inflammatory action. Furthermore, typical coloboma is an hereditary anomaly and nearly always occurs in a certain situation, two facts which are inconsistent with either accidental or inflammatory origin. For certain atypical cases, however, an explanation may be found in prenatal inflammation. Landman reported a case of absence of the choroid in both eyes and attributed the imperfection to embryologic obliteration of the posterior ciliary arteries. Atypical coloboma, *i. e.*, occurring elsewhere than below the nerve, has been explained on the supposition that certain eyes may possess two fetal clefts, a condition which has been observed in some of the lower animals. Notwithstanding our present inability to explain the method in which coloboma of the choroid is formed, its location along the line of the fetal cleft is a potent argument that some fault in the cleft is instrumental in arresting development of the choroid at this place. Microscopic examinations of colobomata of the choroid show that the defect is almost invariably covered by more or less imperfectly formed retina which may consist of all layers or of only a single layer, the membrana limitans interna (Figs. 27-28). Absence of the retina over a coloboma cannot be determined by ophthalmoscopic examination.

STEREOGRAM 8. COLOBOMA OF THE MACULA. Right eyeground of a girl 16 years of age. Left eye normal in all respects. Right eye possesses only peripheral vision. Central light perception cannot be demonstrated. The macular region is occupied by a horizontally oval defect, of about two discs' diameter, evidently a coloboma of the choroid. The fundus contains no other imperfection or evidence of disease. The floor of the coloboma consists of bluish-white sclera, depressed about 0.3 mm. (1.D.) below the plane of the fundus. Dense masses of pigment encircle the defect and cross its surface.

The great majority of choroidal colobomata follow the line of the fetal cleft as shown in stereograms 6-7. Occasionally, however, atypical cases of this malformation occur situated elsewhere than below the disc. Among others, the following examples of atypical coloboma have been reported: coloboma of choroid and iris outward; coloboma of choroid and iris inward; of choroid in one direction and of iris in another; bilateral coloboma outward; coloboma upward, etc. The most common atypical coloboma of the choroid occurs at the macula. In this situation the defect usually is elliptical, the long diameter directed horizontally; rarely it is round, rhomboid or triangular in outline. Its size ranges from two

to six disc diameters. Genuine cases present the appearance of typical coloboma, *i. e.*, the margin is sharply marked and pigmented, the surface usually is depressed and may be crossed by vessels or pigment.

**DIAGNOSIS.** Natal hemorrhages or ruptures of the choroid may result in cicatrices that cannot, with certainty, be differentiated from congenital coloboma. An associated coloboma of the iris is the best evidence that the choroidal defect is a malformation and not a cicatrix. Cases in which the fundus contains other lesions, especially those of chorioretinitis, cannot be accepted as genuine. There is sometimes a tendency for colobomatous eyes to rotate on their anteroposterior axis (torsion); therefore, the position of the eye should be determined before deciding that the direction of a coloboma is atypical.

**STEREOGRAM 9. COLOBOMA OF OPTIC NERVE AND NERVE SHEATH.\***  
Right eyeground of a woman 27 years of age. Vision is 20/50. The ophthalmoscope reveals a colobomatous formation involving the optic nerve. The area of the disc appears greatly enlarged and depressed. The depression is surrounded by a white zone of exposed sclera, level with the surrounding fundus from which it is separated by a ring of black pigment. The pit of the coloboma is shallow above and gradually slopes downward and backward, terminating below in a pouch-like cavity resembling a glaucomatous excavation into which the vessels dip and disappear. Details are indistinct owing to a semitransparent tissue which fills the depression. The surface of the coloboma may be divided into three parts as follows: (1) the nerve head, represented by an oval, pinkish disc, situated above; (2) the surrounding floor and walls of the pit which possess a bluish-white color and represent a defect in the scleral canal and sheaths of the nerve (coloboma of the nerve sheaths); (3) the white zone and pigment ring. The line of pigment marks the limit of the choroid; therefore, the white zone is a defect or coloboma in the choroid. The greatest depth of the coloboma is 1.3 mm. (4.D.).

Several malformations involving the head of the nerve are grouped under the name of "coloboma of the nerve." These are: (*a*) Coloboma of the choroid which includes the nerve; (*b*) Coloboma of the nerve sheaths formed by enlargement of the scleral canal and dilatation of the sheaths. In these cases the sheaths are not closed in front by normal sclera, neither is the coloboma covered by choroid; (*c*) Coloboma limited to the nerve. In fetal life the lower surface of the optic nerve contains a cleft through which the central vessels enter. Failure of this cleft to close constitutes

\*From the service of Dr. David Webster, Manhattan Eye, Ear and Throat Hospital.





**Fig. 27.**



**Fig. 28.**

**Fig. 27—Coloboma of Choroid. The Retina continued over the defect.**  
**Fig. 28—Coloboma of Choroid. The Retina dips into and lines the defect.**



**Fig. 29.**

**Coloboma of Optic Nerve and Sheaths.**

a true coloboma of the nerve. On closure of the fetal cleft, if the retinal vessels are excluded from the nerve they are replaced by ciliary vessels which enter the eye around the pseudo-disc, usually below. Fuchs regards the common inferior crescent of the optic disc (Stergm. 10) as a feebly developed coloboma, a view which has not been universally accepted. The scheme of distribution of the retinal vessels in coloboma of the nerve has been divided by Casper\* into three classes as follows:

(1) All vessels emerge from the lower part of the coloboma or pseudo-disc.

(2) Vessels emerge from near the center of the disc and are distributed in a normal manner.

(3) Vessels enter the eye around the periphery of the coloboma (ciliary vessels).

Colobomata of the nerve and its sheaths present the most varied ophthalmoscopic pictures. The defective area varies in size from two to twenty disc diameters. Usually, the depressed portion is deepest below. Local ectasiæ in the walls of the coloboma are frequently observed. True colobomata limited to the nerve occur as holes in the disc, quite different from a glaucomatous or physiologic excavation. The "punched-out" hole sometimes seen in the disc is regarded by Lindsay Johnson† as a vestigial trait. A similar formation occurs normally in the optic nerve of the elephant. Coloboma of the nerve usually is associated with coloboma of choroid, iris, lens or some other ocular malformation. It is said to exist in 20 per cent of microphthalmic eyes. Vision may be nearly normal; usually, however, it is very defective. Enlargement of the normal blind spot corresponds to the size of the coloboma.

I found the following example of the nerve and sheaths in the eye of an infant who died three weeks after birth:‡ Diameter of the globe was 14 mm. *Microscopic examination.* Anterior hemisphere contains no malformation. Immediately below the head of the optic nerve is a large pit formed by bulging of the dural sheath and external fibers of the sclera. Most of the internal layer of scleral fibers stop abruptly at the edge of the pit. At one side, however, a bundle of fibers continues for a short distance over the pit and then sharply bends forward to form a column of connective tissue (Fig. 29) in the vitreous. Evidently, these strands of connective tissue represent the lamina cribrosa which has made an abortive

\* Casper, Dissertation, Bonn, 1887.

† Lindsay Johnson, Pocket Atlas, etc.

‡ From the laboratory of the New York Post Graduate Medical School and Hospital.

attempt to bridge the chasm. The optic nerve contains the retinal vessels and is well formed except at its ocular end where it dips downward into the colobomatous excavation (Fig. 30). The retina preserves its normal structure until it arrives at the pit into which it enters and forms a tangled meshwork above the nerve fibers. The choroid ends at a short distance from the edge of the pit, leaving a zone of sclera uncovered.

DIAGNOSIS. Coloboma limited to the nerve appears as holes or clefts in the disc, usually situated below. A small coloboma limited to the nerve and sheaths may present a superficial resemblance to glaucomatous excavation. Reference to the following table will show the essential points in which the two conditions differ:

#### COLOBOMA OF OPTIC NERVE AND SHEATHS.

In the absence of congenital malformations, the external aspect of the eye is normal.  
 Eye tension normal.  
 Congenital malformation.  
 Non-progressive.  
 Visual field shows only enlargement of normal blind spot.  
 Usually unilateral.  
 No pain or inflammation.  
 Vessels not diseased but are distributed anomalously.  
 Anatomic landmarks disturbed.  
 Disc varies in color. Lamina cribrosa not seen.

#### GLAUCOMA.

The eye presents the glaucomatous aspect.  
 Tension increased.  
 Disease of adult life.  
 Progressive.  
 Characteristic contraction of nasal field.  
 Both eyes affected though in varying degrees.  
 Even cases of simple glaucoma may, at times, suffer pain.  
 Vessels diseased but are normally distributed.  
 Landmarks undisturbed.  
 Disc grey in color and atrophic.  
 Lamina markings usually distinct.

STEREOGRAM 10. Inferior Optic Crescent (Congenital Crescent; Inferior Staphyloma; Fuchs' Coloboma; Conus nach unten). Right eyeground of a girl 19 years of age. Similar conditions exist in both eyes. Has hyperopic astigmatism. With correcting glasses the vision remains slightly subnormal. The optic disc presents the only abnormality in the fundus. The nerve head is reddened. Adjoining the disc below is a broad, white crescent. The crescent is not depressed and the retinal vessels cross its surface without bending. The pigment which

surrounds the top and sides of the disc is continued around the crescent, so that the latter appears to form part of the nerve head.

The above description is applicable to all well developed cases of congenital, inferior conus or crescent of the disc. A congenital crescent may appear around any portion of the nerve but the great majority are directed downward. In physiologic excavation the cup always opens toward a conus when the two conditions are associated. When a disc with inferior crescent also has a connective tissue ring, the ring and crescent are continuous. As a rule, inferior crescent occurs in eyes with refractive errors and slightly subnormal vision.

Although inferior crescent is a common condition, its anatomic structure is undetermined. Probably, similar ophthalmoscopic pictures are produced by differing conditions. The relation of inferior to temporal (myopic) crescent is uncertain; at all events, they are clinically regarded as independent conditions. Temporal crescent is almost constant in myopia, but inferior crescent is common in hyperopia and hyperopic astigmatism. Fuchs regards the inferior crescent as an incomplete coloboma. If so, it differs from undoubted colobomata in seldom being associated with other formative defects. Elschnig would limit the term "coloboma" to those defects which are depressed below the level of the disc. He regards all crescents as congenital even though they progressively increase in size. Frost states that inferior crescent is an unperforated portion of the lamina cribrosa; in support of which he points out that conus always occurs within the limits and at the expense of the disc. The reddened disc he attributes to crowding of the nerve fibers into insufficient space. Other theories of origin are: (1) exposed sclera; (2) exposed inner surface of nerve sheath; (3) congenital absence of the choroid.

STEREOGRAM 11. OPAQUE NERVE FIBERS (Medullated Nerve Fibers). Left fundus oculi of man, 48 years of age. Similar ophthalmoscopic pictures are presented by the two eyes. Central vision in each eye is normal. The visual fields show enlargement of the physiologic blind spot.

LEFT EYEGROUND. Extending into the retina from the upper and lower margins of the disc are large, white, striate patches terminating in delicate filaments, resembling brush-strokes of white enamel. These patches are typical opaque nerve fibers. Additional fibers are scattered along the inner and outer margins of the nerve and a solitary wisp is observed around the inferior temporal artery. The fibers become opaque first near the periphery of the disc. They pass behind and in front of the



retinal vessels, obscuring or completely hiding them. Although the vessels appear embedded in solid tissue, their circulation is in no way disturbed. The disc appears unduly red, but this is an artefact produced by contrast with the brilliant white nerve fibers.

The nerve fibers which compose the optic nerve have no neurilemma, but each fiber is medullated, *i. e.*, it is enclosed by the fat-like substance known as the myelin sheath. At the lamina cribrosa the nerve fibers lose this myelin sheath and only naked axis cylinders enter the eye. The utility of this change is obvious, inasmuch as medullated fibers are opaque, while non-medullated fibers, designed for the retina, are perfectly transparent. It may happen, as in this case, that certain nerve fibers, after passing into the eye, are again invested with a myelin sheath or the sheath may be continued from the nerve, through the lamina into the retina. In the retina the opaque fibers are ophthalmoscopically visible as brilliant, bluish-white striæ. Opaque nerve fibers are almost constant in the rabbit's retina, and in man they occasionally occur as a physiologic anomaly. Throughout the nervous system development of myelin sheaths proceeds from center to periphery. As they form late in fetal life, medullation of the optic nerve is not completed until a month or two after birth; consequently, opaque nerve fibers in the retina are not, strictly speaking, congenital. Intraocular medullated nerve fibers present a striking ophthalmoscopic picture frequently mistaken by the novice for a pathologic condition. In the great majority of cases opaque fibers are attached to the optic nerve, usually at the upper or lower margin. Very rarely they are confined within the borders of the disc. In other cases the disc and neighboring fundus are normal, while a distant patch of opaque nerve fibers appears on the surface of the otherwise transparent retina. Such isolated fibers usually, but not invariably, appear thin and translucent. The number of opaque nerve fibers varies in different cases from an insignificant patch, which may escape observation, to an enormous white zone completely encircling the disc. When few in number and equally distributed around the nerve they resemble the retinal striation of incipient neuritis.

One or both eyes may be affected, more frequently both. Bilateral cases seldom are symmetrically developed. Blindness exists in those areas occupied by opaque nerve fibers. If fibers are abundant on the macular side of the nerve vision may be reduced, but ordinarily they are so situated as to cause no functional disturbance. The anomaly appears to be hereditary and frequently affects several members of a family.

**DIAGNOSIS.** Patches of opaque nerve fibers disconnected from the disc are especially apt to be mistaken for a manifestation of disease. They are recognized by their filamentous structure, radial direction and by the manner in which they run over and under vessels without affecting the circulation. As distinguished from striations around a choked disc, opaque nerve fibers are sharply circumscribed and brilliantly white: Furthermore, they are not accompanied by swelling of the nerve-head, engorgement of veins, hemorrhage or edema, all of which belong to the clinical picture of choked disc. Opaque nerve fibers are readily differentiated from choroidal disease by their superficial situation. In albuminuric retinitis small, fluffy, striate exudates may occur between the nerve fibers, but, usually, they are accompanied by other exudates and by vascular degeneration. The large "snowbank" exudate which, in albuminuric retinitis, sometimes involves the optic nerve, never presents the compact, white, shining surface of opaque nerve fibers. Retinal light reflexes differ from opaque nerve fibers in that they are widely distributed and they appear and disappear on changing the position of the ophthalmoscopic mirror.

**STEREOGRAM 12. PREPAPILLARY RETINAL ARTERY.** Left eyeground of a boy which, among other physiologic variations, presents a beautiful vascular anomaly. Retinal and choroidal pigment is scanty throughout the fundus. The region around the disc which usually contains an excess of pigment, is almost completely depigmented. A curious "pseudo-conus" exists on both sides of the nerve. The eye is hyperopic and, as is common in this condition, the retinal vessels are rather tortuous. Three vessels which emerge from the margin of the disc, hook over its edge in a manner characteristic of cilioretinal vessels. The most striking feature in the fundus is a vascular loop, twisted like a rope, which projects from the disc into the vitreous for a distance of 2.6 mm. (8.D.). It originates close to the central vein but in color resembles an artery. The base of the loop is enclosed in connective tissue. The loop undergoes strong, systolic pulsations during which its anterior extremity rises in the vitreous and the entire coil appears to elongate.

Although the distribution of retinal arteries conforms to a general plan, the widest variations in their arrangement exist upon the disc. Frequently an artery and vein entwine, a vein is twisted about an artery (Sternberg, 11-33), or a vessel may coil upon itself. Vessels may leave the nerve, form a loop and re-enter it again. Hirschberg reported a case in which a prepapillary artery entered the vitreous where it broke into numerous branches which entered the peripheral parts of the retina.

**DIAGNOSIS.** Preretinal vessels usually are confounded with the persistent hyaloid artery from fetal life. Examples of the two anomalies which I have seen present essential differences. The preretinal artery is a well formed vessel which, after its excursion into the vitreous, returns to the nerve. This vessel presents every appearance of having always pursued its present course. The hyaloid artery, on the contrary, is primarily distributed to the fetal lens and after incomplete absorption it does not possess the finished circulatory connections of a preapillary loop. So-called, persistent hyaloid arteries consist principally of membranous remnants of the hyaloid vascular sheath, the artery itself frequently being absent or forming but an insignificant part of the structure.

**STEREOGRAM 13. PSEUDONEURITIS AND TORTUOUS VESSELS IN HYPERMETROPIA.** Right eyeground of healthy man, 27 years of age. Has a high degree of hypermetropia in both eyes which present similar ophthalmoscopic pictures. With correcting glasses the vision is normal. The fundus is darkly stippled. The nerve-head is uniformly reddened and elevated about 0.66 mm. (2.D.). By the direct method of examination the margin of the disc is indistinct but by the indirect method the outlines become sharp. The retinal vessels, both arteries and veins, are very tortuous but the arteries are not disproportionately narrower than the veins. All vessels are uniformly broadened and present neither local constrictions nor dilations. There are no hemorrhages or exudates in the retina. Prolonged observation showed the fundus condition to be stationary. Diagnosis is hyperopic fundus or pseudoneuritis.

The term "pseudoneuritis" is applied to a congenital condition which beginners in ophthalmoscopy almost invariably mistake for optic neuritis. Nearly all typical examples occur in highly hyperopic eyes, *i. e.*, eyes below the normal size. The disc is dark red in color and fine supernumerary vessels can be discerned on its surface. The outline of the nerve is more or less blurred and frequently covered with delicate striæ, especially on the nasal side where nerve fibers are most abundant. Sometimes the nerve is surrounded by a delicate grey halo. The head of the nerve may project 1-3.D. thus increasing the resemblance to papillitis. The disc may be unusual in form or contain an inferior crescent. The retinal vessels are unusually tortuous and on the disc they may twist, form loops, or divide in an erratic manner. The appearance of neuritis presented by highly hyperopic eyes is due to crowding a normal sized retina into an undersized scleral cavity. As a result the retinal vessels adapt themselves to insufficient space by becoming tortuous, the





**Fig. 30.**  
**Coloboma of the Optic Nerve and Sheaths.**



**Fig. 31.**  
**Pseudoneuritis.**  
**Congenitally small eye. Retina crowded into an undersized Scleral Cavity.**

nerve head, overcrowded with capillaries, is reddened, and the nerve fibers, unable to extend their full length, are piled up on the disc. In hyperopia the disc is redder than in emmetropia, but only about five per cent of cases exhibit pronounced pseudoneuritis. Pseudoneuritis is a stationary condition, differing in this respect from neuritis which either recedes or becomes worse.

In all the congenitally very small eyes which I have microscopically examined the retina was disproportionately large. The exaggerated example shown in Fig. 31 is from a child two years of age. Polar diameter of the globe was 18 mm.

The appearance of mild neuritis may be simulated also by glassy, radiating fibers which surround the disc and obscure its outline. This picture is produced by semiopaque nerve fibers and, if careful search is made, it is usual to find a few densely opaque fibers in one or the other eye. A deceptive appearance of commencing edema of the nerve may be produced by physiologic light reflexes. Neither of the two last mentioned conditions is accompanied by tortuous vessels. Slight opacities in the cornea, aqueous, lens or vitreous will obscure and darken the disc producing an effect frequently mistaken for congestion.

**DIAGNOSIS.** Both active and passive hyperemia of the disc occur but, owing to the wide variations in color presented by normal discs, the condition is extremely difficult to diagnosticate. Mere redness of the nerve, however marked, does not warrant a diagnosis of inflammatory hyperemia unless some of the following conditions coexist:

(a) Hyperemia greater than previously existed in the affected eye or now exists in the fellow eye.

(b) Redness which extends into a physiologic excavation.

(c) The outline of the disc appears blurred by both direct and indirect methods of examination.

(d) Small vessels on and around the disc increased in numbers.

(e) Veins enlarged and tortuous with arteries straight and narrowed.

(f) Individual vessels exhibit inequalities in breadth.

(g) Lymph stasis in perivascular sheaths manifest by light lines along vessels, on and near the disc.

(h) Absence of high hypermetropia and astigmatism.

True neuritis does not commence with diffuse redness, but with distinct edema and blurring of the nasal side of the disc and adjacent retina (Stergm. 66). As stated by Frost, congenitally tortuous vessels do not usually rise above the level of the fundus, while in pathologic tortuosity

the sinuosities of the vessels are perpendicular to the plane of the retina. Pseudoneuritis is differentiated from true neuritis, chiefly, by absence of all pathologic symptoms and presence of hypermetropia or astigmatism. Crowding of the retina, however, is not due to refractive error but to the small size of the eye and small eyes are not always hyperopic or astigmatic.

True hyperemia of the disc may accompany choroiditis, retinitis, diabetes, nephritis, high blood pressure, polycythemia, retrobulbar neuritis, nasal sinus disease, brain lesions, certain diseases of the central nervous system, cerebral sinus thrombosis, cardiac disease, mania, etc. It may follow poisoning by certain drugs especially lead, iodiform and alcohol.

STEREOGRAM 14. PERSISTENT HYALOID ARTERY. PERSISTENT CANAL OF CLOQUET. Right eyeground of a girl 13 years of age. Left eye normal. The only abnormality observed in the right eye is a bluish-white cord extending from the center of the disc far into the vitreous, terminating in a number of filaments (not shown in picture). Its anterior extremity is seen best with a plus lens of 16.D. The slightest movement of the eye causes active oscillatory motions in the cord, but its excursions are of limited extent. No blood vessels are visible on its surface. Evidently, the formation is a congenital anomaly, clinically known as persistent hyaloid artery.

In the third month of fetal life the central artery of the retina, enclosed in a cellular sheath, extends through the vitreous to the posterior surface of the lens which it invests with a vascular covering. The vessel, in its course through the vitreous, follows the hyaloid (Cloquet's) canal and is named the hyaloid artery. By the end of the ninth month the hyaloid artery should be completely absorbed. Occasionally the artery or sheath is not entirely removed, in which case they persist as fetal vestiges. These remnants of intrauterine life show the widest variations in structure. They may remain attached to either the nerve, the lens or to both. The most common example of incomplete absorption is an opaque patch on the posterior capsule of the lens (posterior polar cataract) which may or may not be accompanied by a tag of tissue on the nerve. A persistent hyaloid artery contains tissues from both the artery and its sheath. When the artery persists it may be single or divided into several branches. A common formation is a cord or membrane representing either the obliterated artery or the cellular sheath. These structures vary in form from a thin filamentous thread to a solid column of connective tissue in which blood vessels are insignificant or absent. Great diversity exists in

the anterior termination of the cord or membrane; thus, it may be attached to the lens, divide into branches, split into fine filaments, terminate in a bulb or in a sharp point, etc.

**DIAGNOSIS.** It is not certain that all the membranous and vascular structures in the vitreous, clinically known as persistent hyaloid arteries are rightly interpreted. Eversbusch\* does not regard differentiation as easy, and separates these formations into two classes: (1) True persistent hyaloid artery; (2) Membranous opacities in the hyaloid canal, probably the residuum of a hemorrhage. v. Reuss† agrees with Eversbusch and for the diagnosis of persistent hyaloid artery requires that the vessel should distinctly arise from the arteria centralis retinae and be filled with blood. Greeff‡ diagnosticates persistent hyaloid artery when the cord is connected with the central artery and posterior pole of the lens or else the anterior end terminates in numerous branches. The cord should not exceed the calibre of the central artery. Greeff further diagnosticates a visible Cloquet's canal when the formation has a funnel or ampullaform shape and the contents are opaque. A persistent visible canal may contain embryonic tissue or a well developed artery. After partial absorption of vitreous exudates I have seen opacities persist along the course of Cloquet's canal which closely resembled persistent fetal elements. Such cases are easily differentiated by the history of fluctuations in visual power which do not occur with fixed congenital defects. The membranes of retinitis proliferans usually are too widely distributed to be mistaken for fetal remnants of the hyaloid canal. Nevertheless, after injury with intraocular hemorrhage, membranes may form which, in the absence of a history, could be mistaken for persistent, visible hyaloid canal (Fig. 138). A pre-papillary artery (Stergm. 12) is frequently miscalled persistent hyaloid. It is distinguished by its completed structure, perfect circulation and by its origin and termination in the nerve.

**STEREOGRAM 15. ALBINISM.** Eyeground of an albino girl 14 years of age. The ophthalmoscopic picture is that of an exaggerated blond fundus in which the normal pigment of the eye is absent. The background of the fundus has a pinkish-white color produced by the white sclera shining through the capillary layer of the choroid. The optic disc is of normal color but appears dark from contrast with the white fundus. The blood vessels form a beautiful red tracery against the pink-white sclera. The retinal vessels lie in front of the choroid and are recognized by their

\* Eversbusch, Mitt. a. d. Klin., München, 1882.

† v. Reuss, Mitt. d. ii Wien. Klin., ii, 1885.

‡ Greeff, Orth's Sp. Path. Anat. ii Half. ii, p. 556.



rounded form and terminal manner of distribution. The choroidal vessels are sharply defined against the light background. They have a ribbon-like appearance, possess no central light streak and anastomose freely with each other. The deeper red color of the macula suggests slight pigmentation of that region.

Absence of the physiologic pigment in the body is termed albinism. The anomaly is most common among the dark skinned races. Albinism may be complete or incomplete. It would appear that pigment is lacking in the eye only in complete cases, and it is not uncommon to find a well pigmented fundus in the eye of an individual whose hair and eyelashes are nearly albinotic. Pigmentation of the choroid does not occur until shortly before, or even after, birth; consequently, infants' eyes frequently contain islands of unpigmented choroid, particularly at the periphery. Occasionally, these unpigmented areas persist through life. Late pigmentation of the choroid explains why some cases of albinism improve during the first years of life. In complete congenital albinism pigment has not been deposited in the skin, hair, eyelashes, eyebrows, uveal tract or retinal epithelium.\* The hair is fine and downy. The iris is a light grey color but becomes pink when the eye is transilluminated. The pupil has a deep red color: It is usually contracted but is very active. The albinotic iris being translucent, fails to perform its normal function of diaphragm to camera, and these unfortunates are almost blinded by the excess of light which enters the eye. In their efforts to escape the distressing illumination the eyelids are spasmodically compressed and the eyes rotate from side to side until permanent nystagmus is established. Myopia and astigmatism usually develop, probably produced by pressure and traction exerted on the globe by spasm of the extrinsic muscles. The cells of the retina and choroid are normal aside from absence of pigment. Vision usually is subnormal, but the color sense is unaffected. Naturally, vision is better at dusk than in daylight. No cause for the anomaly is known. Consanguinity does not appear to be influential in its production. It is hereditary and may occur in several members of a childship.

**DIAGNOSIS.** Congenital albinism of the fundus associated with general achromatism is almost unmistakable. Pathologic depigmentation of the eyeground, however, does occur and the ophthalmoscopic picture which

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\* It is stated, that in human albinos the retinal epithelium always contains pigment. This requires confirmation, inasmuch as no pigment is contained in the retinal epithelium of albino rabbits.

results closely resembles that of the albinotic eye (Stergm. 26). The only case I ever saw occurred in a diabetic. The differential points of this case are contained in the following table:

| CONGENITAL ALBINISM OF<br>EYEGROUND.                                                                                                                                                                                                                                           | ACQUIRED ALBINISM OF<br>EYEGROUND.                                                                                                                                                                                                                                                         |
|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <p><b>Congenital.</b><br/> Associated albinism of hair, iris, etc.<br/> Photophobia; under proper conditions vision and fields, practically, normal.<br/> Vessels of choroid normal.<br/> Depigmentation of fundus uniform and complete.<br/> No systemic disease present.</p> | <p><b>Acquired.</b><br/> Not associated with general albinism.<br/> No photophobia, but serious impairment of vision and fields.<br/> Many choroidal vessels atrophic.<br/> Depigmentation irregular. Fundus contains scattered specs of pigment.<br/> Affected with systemic disease.</p> |

**STEREOGRAM 16. PUNCTATE CONDITION OF THE FUNDUS (Gunn's Dots, Crick Dots).** Right eyeground of a healthy housemaid 20 years of age. Fundus conditions are identical in both eyes. With correcting glasses vision is normal. The macula and surrounding region contain numerous groups of very pale dots, situated on the surface of the retina. The nearer to the fovea, the more distinct are the dots.

The title "punctate conditions of the fundus" was applied by Frost\* to a group of varied conditions, some of which have since been more definitely classified, while the nature of others, however, remains unknown. Chief among the former is a dotted condition of the macular region, first described by Marcus Gunn and since known as Gunn's dots or, from the family name of Gunn's first cases, "Crick dots." These terms are usually applied to small groups of light colored spots, situated chiefly at the fovea. They appear to be superficial and never exceed in size the diameter of a medium sized retinal vessel. These dots are inconspicuous and easily overlooked. They are seen best by weak illumination with the light directed obliquely. The color is described as white or yellowish. All those which I have observed are the color of the eyeground but very much paler. Gunn's dots occur in perfectly normal eyes and are without pathologic significance. They are most distinct in early life when all the refracting media of the

\* Frost, The Fundus Oculi, 1896.

eye are perfectly transparent. The fundus stereogram here shown is remarkable for the excessive number of dots, but in all other respects the case corresponds with Gunn's description.

The anatomy of Gunn's dots is unknown. They are commonly attributed to inequalities on the surface of the retina which reflect light. In well developed cases it often happens that the retina around the dots is stippled, an appearance presumably due to rarefaction of the epithelial pigment. In the case shown in the stereogram the appearance suggested that the dots and stippling were alike produced by a sieve-like reticulation of the pigmented epithelium.

**DIAGNOSIS.** A diagnosis of "Gunn's dots" should be made only in eyes that are perfectly normal; never where deterioration of vision has occurred. Central choroiditis is distinguished by the greater size and distinctness of the spots and by visual disturbance. Colloid excrescences (drusen) usually occur in the aged. They are more widely distributed and more distinct than Gunn's dots. Minute, isolated glistening specks are sometimes seen in the normal fundus. They occur at all ages and in any part of the eyeground. Their brilliant metallic luster is sufficient to differentiate them.





Fig. 32.

Suppurative Choroiditis.

Retina detached by Purulent Exudates from Retina and Choroid.



Fig. 33.

Choroiditis.

Stage of Exudation.





Fig. 34—Choroiditis,  
Stage of Exudation.

Fig. 34.

Fig. 35—Choroiditis,  
Formation of Granulation  
Tissue which in  
this case is very  
abundant.



Fig. 35.







Fig. 36—Choroiditis Stage of Cicatrization.

Fig. 36.



Fig. 37—Choroiditis, advanced stage of Cicatrization. Proliferation of Pigment Cells.



Fig. 38—Choroiditis Hyperplastica.

Fig. 38.

## CHAPTER IV.

### AFFECTIONS OF THE CHOROID.

Inflammation of the choroid is broadly classed as suppurative (acute) and exudative (chronic) choroiditis. Suppurative choroiditis is an acute infectious process. Owing to the early and rapid development of vitreous opacities the fundus changes cannot be seen with the ophthalmoscope (Fig. 32).

**EXUDATIVE CHOROIDITIS.** This term includes the various types recognizable with the ophthalmoscope. Inasmuch as the retina participates in the pathologic process, the disease is also termed *chorioretinitis*. The clinical forms of exudative choroiditis are named according to the situation, arrangement or form of the lesions as follows: c.centralis; c.anterior; c.disseminata; c.diffusa; c.circinata; c.areolaris; c.guttata, etc. Many of these terms are merely descriptive of special cases, not special forms of disease. The majority of cases present scattered foci of disease which subsequently may fuse and form large patches. A diffuse exudative inflammation of the choroid is uncommon.

**PATHOLOGY.** Individual lesions of a choroiditis pass through three stages, (1) exudation (Figs. 33-34); (2) formation of granulation tissue (Fig. 35); (3) cicatrization (Figs. 36-37). With the ophthalmoscope it is not practicable to differentiate the stage of exudation from that of granulation. In the exudative stage foci of round cells accumulate about the blood vessels of the inner layers of the choroid. The pigmented stroma cells of the choroid emigrate inward and appear in the inner layer where they are never found under normal conditions (Figs. 33-34). At the same time a fluid exudate, which varies greatly in amount, infiltrates the choroid and separates it from the retina (Fig. 34). In the second stage the amount of granulation tissue varies. Usually it is slight but may be abundant (Fig. 35). In certain cases it may invade the vitreous (choroiditis hyperplastica) (Fig. 38). The retinal epithelium, which is nourished by the choroid, being deprived of nutrition exfoliates and parts with its pigment which may appear on the surface of the retina in the form of specks or branching figures.

**STEREOGRAM 17. CHOROIDITIS DISSEMINATA, EXUDATIVE STAGE**

(Eruptive Stage). Right fundus oculi of a woman 32 years of age. No history of either syphilis or tuberculosis. Both eyes are equally affected. Vision 20/40. Flashes of light and a "glimmering" sensation are experienced. The ophthalmoscopic picture is that of early choroiditis disseminata. The fundus, especially the equatorial region, is covered by an eruption of discrete, round, softly-outlined, reddish-yellow spots, situated below the level of the retinal vessels. The spots vary in size from one-fourth the diameter of the disc to that of a primary retinal vessel. At certain points they occur in thickly crowded clusters. In this early stage of the disease the spots show but slight tendency to coalesce and their borders are not pigmented. Two older cicatricial spots are seen, bordered by pigment. Although the patient is a brunette, the entire fundus is light in color and the choroidal vessels are plainly seen, indicating slight, general depigmentation of the retinal epithelium. The retina is transparent and the vessels normal. The optic disc is reddened, presumably due to hyperemia of the choroid which has extended to the head of the nerve through the anastomosing vessels of Zinn.

A slight knowledge of the histologic changes described enables one to interpret the picture shown in the stereogram. For example, the flattened lesions and transparent retina indicate that the exudate is slight in amount and the granulation tissue not exuberant. The color and distinctness of the spots are modified by the amount of pigment remaining in the overlying retinal epithelium; thus, where the pigment is not disturbed the lesion is obscure, but where the pigment has been partially discharged the spots are grey, while those spots over which depigmentation is complete possess a reddish-yellow color produced by the red choroid seen through the yellow exudate. Inflammatory products from the choroid do not readily pass through the lamina vitrea into the vitreous unless the exudate is very abundant or the lamina perforated. As a rule, the vitreous is clear in choroiditis of the posterior and equatorial regions. When, however, the disease develops far forward or involves the ciliary body, the anterior part of the vitreous is filled with floating opacities (Fig. 39).

Exudative choroiditis is caused by a variety of general infections, especially syphilis and tuberculosis. Other causes are traumatism, local infection and, probably intestinal autointoxication. In many cases of the disseminate form the etiology is obscure. The course of disseminated choroiditis is slow. Usually the prognosis is unfavorable. In certain cases, however, the progress of the disease is arrested, but the diseased

areas cicatrize and remain as permanent gaps in the visual field. Choroidal lesions at the macula seriously impair vision, but situated at the periphery they may not be troublesome.

**DIAGNOSIS.** Choroiditis in the exudative stage requires differentiation chiefly from angiopathic retinitis of albuminuric or diabetic origin. In choroiditis the spots are yellowish in color, deeply situated below the level of the retinal vessels and some pigmentation is usually present. In

|                                        | EXUDATIVE<br>STAGE OF<br>CHOROIDITIS<br>DISSEMINATA                                                 | EXUDATIVE<br>STAGE OF<br>ANGIOPATHIC<br>ALBUMINURIC<br>RETINITIS                                                                | TYPICAL CASES<br>OF CENTRAL<br>PUNCTATE<br>DIABETIC<br>RETINITIS                                                                        |
|----------------------------------------|-----------------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------------------------------------------------|
| Number and<br>Situation<br>of Lesions. | Numerous and widely distributed but favor equatorial region. Always below level of retinal vessels. | Comparatively few in number. Favorite site is posterior polar zone. Superficially situated and often cover the retinal vessels. | Numerous. Occur chiefly in macular region. Superficially situated and cover retinal vessels.                                            |
| Form and<br>Color.                     | Yellowish; circular with tendency to coalesce and form scalloped patches.                           | Whitish, "frosty" color. Circular with no tendency to coalesce.                                                                 | Whitish, round and linear spots which do not coalesce.                                                                                  |
| Pigmentation.                          | A few pigmented, cicatricial spots usually present. Fresh spots may be "peppered."                  | No disturbance of pigment.                                                                                                      | No pigmentation.                                                                                                                        |
| Vessels.                               | Retinal arteriosclerosis forms no part of choroiditis disseminata.                                  | Arteriosclerosis usually is a marked feature.                                                                                   | Visible vascular changes are unusual.                                                                                                   |
| Hemorrhage.                            | No retinal hemorrhage in uncomplicated cases.                                                       | Retinal hemorrhages usually present.                                                                                            | Numerous small, retinal hemorrhages between spots.                                                                                      |
| Optic Nerve.                           | Disc may be reddened but there is no neuritis. Retinal edema may extend to nerve (Sergm. 18).       | More or less papillitis usually present.                                                                                        | Neuritis is unusual in this form of retinitis. Diabetic retinitis frequently indistinguishable from other forms of angiopathic disease. |

Retinitis punctata albens, senile "drusen" and Gunn's dots are noninflammatory punctate conditions not apt to be mistaken for any of the above affections.

retinitis, on the contrary, the spots have a whitish "frosty" color, are superficially situated, often overlying the retinal vessels and, in the early stage, are never accompanied by pigmentation. The lesions of choroiditis are numerous and widely distributed while those of retinitis are comparatively few in number and usually appear around the posterior pole. Vascular degeneration, retinal edema and hemorrhage which belong to the picture of angiopathic retinitis are exceptional events in choroiditis. The disc may be reddened in choroiditis, but actual neuritis, so common in albuminuric retinitis does not usually occur. The subjective symptoms of choroiditis, *i. e.*, photopsiæ, metamorphopsiæ and chromotopsiæ are unusual in angiopathic retinitis.

Central diabetic retinitis (Stergm. 38) differs from choroiditis in the following respects. It is confined chiefly to the macular region and is accompanied by hemorrhage; the spots do not fuse and none of them are pigmented nor do they undergo cicatrization.

Retinitis punctata albescens (Stergm. 54) is easily distinguished by the atrophic condition of the nerve and retina; also, by the subjective symptoms of night blindness and contracted fields.

STEREOGRAM 18. CHOROIDITIS DISSEMINATA, EXUDATIVE STAGE. Right eyeground of a girl 15 years of age. Hereditary syphilitic. Has saddle nose, notched incisors, scars at angles of mouth and arborescent corneal opacities characteristic of a preceding interstitial keratitis. Both eyes affected. The left eyeground is shown in stereogram 20. *Right Eye*:—Central vision 20/40. Has subjective visual sensations consisting of flashes of light, balls of fire, etc. Unable to see in a dim light (torpor retinæ). The entire fundus is covered with spots and plaques of disseminated choroiditis. Most of the lesions shown in the picture are in the stages of exudation or granulation, but those in the anterior zone of the fundus (not shown) are in the cicatricial stage. The older spots point to a preceding attack of choroiditis which, probably, developed in connection with a keratitis which occurred six years ago. The field shown in the stereogram is covered with yellowish, softly outlined spots. Originally, all the spots were round, but many of them have coalesced and formed large patches with scalloped edges. Some of the spots already show commencing marginal pigmentation. All the lesions are deeply situated and, when in contact with the retinal vessels, the latter pass over them. Two old, well pigmented spots are seen on the temporal side. On the nasal side of the disc is a large circular area of opaque retina. The vessels which curve over its surface, measured with the ophthalmoscope, are about 0.5



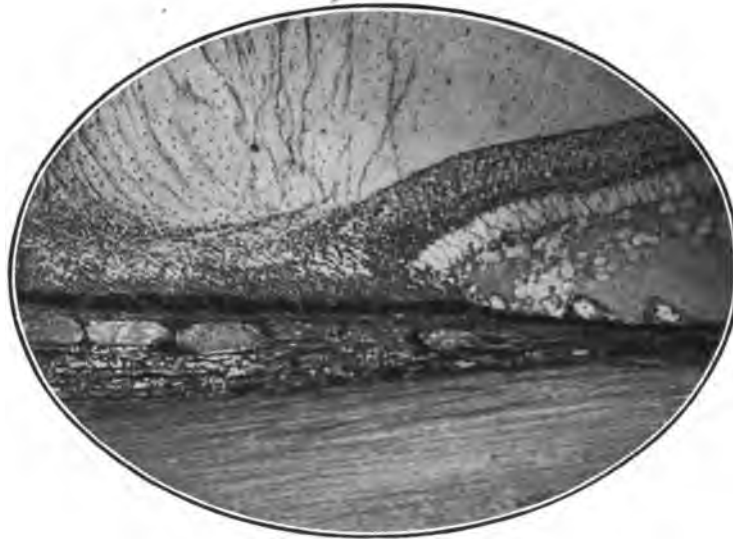


**Fig. 39.**

**Choroiditis.**

**Vitreous opacities are shown in the upper part of the figure.**

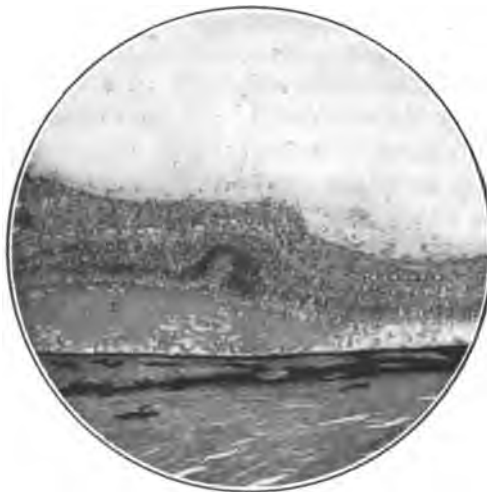


**Fig. 40.**

**Choroiditis causing flat Retinal Detachment.**

**An exudation from the choroid at E has lifted and infiltrated the retina.**

mm. above the level of the disc. This opaque area is a flat retinal detachment produced by a recent, profuse exudation from the choroid which has lifted and infiltrated the retina (Fig. 40). A larger amount of exudate would have passed through the retina and appeared in the vitreous. The infiltration of the retina has extended along the nerve fibers to the disc and obscured its outline on the nasal side. This condition of the nerve-head is distinguished from true neuritis by its evident connection with adjacent edematous retina, also by the absence of swelling and by the normal retinal vessels.\* The disease is a wide-spread inflammation of the choroid manifest by general disturbance of choroid and retina throughout the affected region. In many places the surface of the retina contains a dust-like pigment which has been discharged from the retinal epithelium. The entire eyeground is mottled, suggesting foci of exudation in the deep layers of the choroid.

**SYMPTOMS.** The early subjective symptoms of exudative choroiditis are fairly uniform in all cases. Photopsiæ occur as glimmering, flashes of light, etc., due to disturbance of the rods and cones in the retina. Another symptom is *micropsiæ*, i. e., objects appear too small. This phenomenon is due to stretching of the retina and separation of its percipient elements at the macula by exudate. From the same cause objects may appear distorted (*metamorphopsiæ*). In other cases the macular structures are crowded together and objects appear too large (*macropsiæ*). Visual acuity is always blunted in disseminated choroiditis, but the loss cannot be estimated from the ophthalmoscopic picture. Fair central vision may exist in connection with wide-spread destruction of the fundus. On the other hand, vision may be greatly reduced with but slight visible change. A small central lesion may render the eye useless, while a large peripheral patch may not be noticed by the patient. Torpor retinæ usually exists, and the patient sees well only in bright daylight (hemeralopia). Exceptionally, the retina is hypersensitive and vision is best at dusk (nyctalopia). There may be acute exacerbations in the course of a chronic choroiditis. In the case of stereogram 18 a preceding attack was accompanied by interstitial keratitis, the choroiditis being confined to the anterior zone of the fundus. Such cases are common in congenital syphilis. The keratitis masks the choroiditis and the latter is not discovered until the cornea clears.

**DIAGNOSIS.** In most cases, the presence of old choroidal cicatrices renders it easy to recognize the primary seat of the disease. In rare cases

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\* A somewhat similar condition of the disc is shown in stereogram 59.



chorioretinitis commences by a large subretinal effusion which produces an opaque patch in the retina, similar to that seen on the nasal side of the disc in the stereogram. In such cases the ophthalmoscopic picture is not unlike that which follows closure of a branch retinal artery (Stergm. 45). The two conditions are differentiated as follows:

| OPAQUE RETINA DUE TO<br>CHOROIDAL EFFUSION.                                                                                                                                                         | OPAQUE RETINA DUE TO<br>BRANCH CLOSURE.                                                                                                                                                                                   |
|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Surface elevated.<br>Outline of opacity bears no relation to distribution of artery.<br>Vessels normal or dilated.<br>Gradual failure of vision.<br>Photopsiæ.<br>Usually some pigment disturbance. | Surface not elevated.<br>Fan-shaped opacity corresponding to distribution of branch artery.<br>Arteriosclerotic changes in artery.<br>Sudden loss of vision in affected area.<br>No photopsiæ.<br>No pigment disturbance. |

STEREOGRAM 19. CENTRAL EXUDATIVE CHOROIDITIS. EXUDATIVE STAGE. (Syphilitic). Right eyeground of a man 30 years of age. Wassermann blood reaction for syphilis is positive. Acquired chancre ten months ago. Both eyes are uniformly affected. *Right eye*:—Vision in full daylight is 20/70, but in a dimly lighted room it is but 10/200. He sees flashes of light and shooting stars. Objects at which he looks appear to quiver. The anterior part of the vitreous contains fine, dust-like opacities, seen best by weak illumination and with a strong convex lens in the ophthalmoscope (vitreous opacities not shown in the stereogram). Dust-like vitreous opacities combined with choroiditis are regarded as pathognomonic of syphilis. The optic nerve appears reddened and its margin blurred, appearances partly due to opacities in the vitreous.\* The retinal vessels appear normal. The macular region contains a group of small, rounded spots which show a tendency to coalesce. The older spots occupy the center of the group. They are whitish in color and sprinkled with fine pigment. The more recent spots, situated near the periphery of the group, are softly outlined, reddish-yellow in color and nonpigmented. The normal macular light reflexes are abolished, due to swelling or exudation in the macular depression.

\* The diagnosis of "slight neuritis," "red nerve," etc., should never be made until it is certain that the obscure appearance of the disc is not an artefact caused by opacities in the cornea, aqueous, lens or vitreous.

The term "choroiditis centralis" has been loosely applied to several conditions which differ essentially in etiology and pathology. These are, (a) choroiditis of syphilitic, tubercular or uncertain origin (Stergm. 19-23); (b) atrophy and degeneration of the retina and choroid in myopia (Stergm. 71); (c) senile changes at the macula (Stergm. 27); (d) results of blunt force, hemorrhage, etc. (Stergm. 59); (e) congenital anomalies (Stergm. 8).

The term should be reserved for cases included in class (a).

When choroiditis of syphilitic origin appears in the macular region the eruption may appear as a large, single exudate or as a circumscribed group of closely aggregated spots resembling those of choroiditis disseminata. The symptoms described in the above case are typical of the affection. The subjective symptoms of photopsiæ, quivering of objects, distortion, etc., are due to disturbance of the outer retinal layer. The anterior, dust-like vitreous opacities, usually present in early syphilitic cases, come either from the anterior extremity of the choroid or posterior extremity of the ciliary body. If the ciliary body is extensively affected (cyclitis) the exudate will be carried forward by the aqueous current and deposited, in the form of fine flecks, on the lens and cornea.\* Exudates in the fore part of the vitreous or in the post lental region may be resorbed with astonishing rapidity, while exudates deep in the vitreous disappear slowly and often incompletely.

DIAGNOSIS. Aside from positive evidence secured by the history and Wassermann test, syphilis is indicated in this case by the central location of the eruption and vitreous opacities.

Colloid excrescences (drusen) (Stergm. 27) differ from a central choroiditis as follows. They indicate a degenerative process, usually senile, are nonpigmented and cause no disturbance of vision.

The ophthalmoscopic features of central choroiditis differ from Gunn's dots (Stergm. 16) in the following respects. The choroidal lesions are more distinct, more deeply situated and attended by pigment disturbance. Vision is affected by central choroiditis but not by Gunn's dots.

STEREOGRAM 20. CHOROIDITIS DISSEMINATA, CICATRICIAL STAGE. Left eyeground of girl, 15 years of age. (Right eyeground shown in

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\* Cyclitic deposits in the anterior chamber were long misunderstood. Hence the misnomers "Descemetitis," "serous iritis," etc., formerly employed.

Stereogram 18). Has all the stigmata of congenital syphilis, *i. e.*, saddle nose, notched incisor teeth, cicatrices at angle of mouth and scars of an old interstitial keratitis. Left eye possesses only bare perception of light. Patient is uncertain when vision was lost but thinks it occurred during an attack of keratitis six years ago. The vitreous is clear as is usual in late choroiditis disseminata. The entire fundus is covered with the atrophic lesions of exudative choroiditis. The disease has run its course and after destruction of the choroid and outer layers of the retina, the havoc has been repaired by formation of scar tissue. Complete atrophy of the choroid exists in the broad, white zone about the optic disc, where nothing remains of the vascular structure. Extending outwards from the disc into the macular region, is a broad, dirty-brown tract which, probably, marks the site of a diffuse, subretinal exudation, similar to that existing in the left eye (Stergm. 18). Enormous proliferation of pigment has occurred throughout the entire fundus, especially around the cicatrices. The active proliferation and massing of pigment at the border of a choroidal cicatrix mark the futile efforts of the pigmented epithelium to cover over the defect. Notwithstanding extensive destruction of the choroid and outer layers of the retina, the optic nerve and retinal vessels are nearly normal, indicating that the inner layers of the retina have escaped atrophy. The outline of the disc is blurred due to absence of the choroid which, normally, sharply outlines the nerve. Only the large choroidal vessels remain in the atrophic patches, the capillaries and vessels of medium size having been destroyed.

REPAIR. In exudative choroiditis repair is effected by formation of granulation tissue which is replaced by a fibrous cicatrix that firmly unites the retina and choroid (Fig. 41). The choroid and outer layer of the retina undergo atrophy and form a thin, translucent membrane (Fig. 42), through which the white sclera is visible. The pigmented cells of the choroid (chromatophores) usually perish, but their pigment is transported by leucocytes and deposited around the scar. The retinal epithelium over the lesion is destroyed, but the surrounding epithelial cells undergo active proliferation, covering the granulating surface and, in the final stage, depositing great masses of pigment on and around the cicatrices (Fig. 43). Exudates or hemorrhage in the suprachoroidea (Fig. 44) frequently organize and form an adventitious membrane outside the choroid (Figs. 45-46). When fully organized these suprachoroideal products of inflammation have been mistaken for fibromata (Fig. 47). The structure and



Fig. 41.

**Choroiditis, Stage of Repair.**

A fibrous cicatrix at a, unites the retina and choroid.

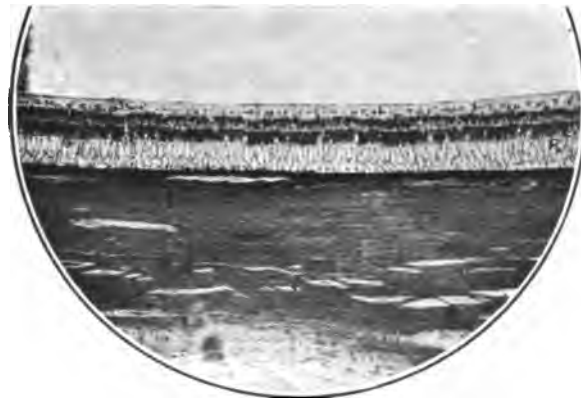


Fig. 42.

**Choroiditis, Atrophic Stage.**

R, Retina; S, Sclera. The choroid is markedly thin and atrophic.





**Fig. 43.**

**Choroiditis, Final Stage of Atrophy.**

**Masses of pigment are deposited on and around the cicatrices.**



**Fig. 44.**

**Exudates in the Suprachoroid.**

**The fibers of the suprachoroid are separated and distended by the effusion, shown in the section at a.**





Fig. 45.

**Choroiditis, Atrophic Stage.**

**Formation of adventitious membrane outside the choroid from organization of exudates and hemorrhage. M, membrane; B blood; S, sclera.**



Fig. 46.



Fig. 47.

**Fig. 46—Choroiditis. Atrophic stage. C, choroid; M, adventitious membrane; S, sclera. Fig. 47—Choroiditis. Organized connective tissue of the suprachoroid, resembling fibromata.**









**Fig. 48.**

**Choroiditis, Stage of Repair.**

**The Choroid is partially replaced by cicatricial tissue. The eye tension has remained normal.**



**Fig. 49.**

**Choroiditis, Stage of Repair.**

**Membranous cicatrices have formed in the Choroid.**





Fig. 50.

Fig. 50—Formation of irregular masses of Fibrous Tissue in the Choroid of a Phthisical Eye.

Fig. 51—Formation of Fibrous Tissue in the Choroid.

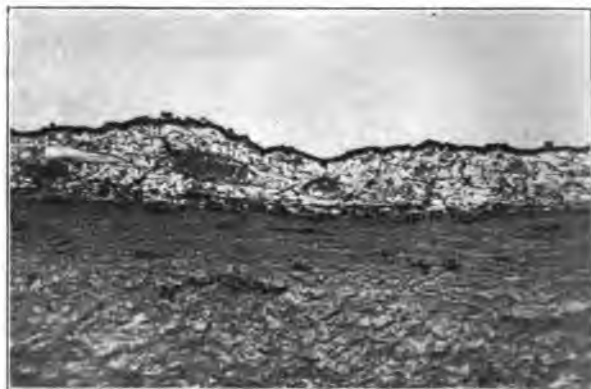


Fig. 51.



Fig. 52.

Fig. 52—Calcification of the Choroid.

amount of cicatricial tissue that replaces the choroid are modified by intraocular pressure. If the eye tension remains normal, or is increased, the cicatrix is membranous or composed of strands of connective tissue (Figs. 48-49). When, however, the eye softens and becomes phthisical, thick, irregular masses of fibrous tissue develop (Figs. 50-51). These hypertrophic fibrous masses may subsequently be converted into bone (Fig. 52).

Unfortunately, the most complete regeneration attainable after choroiditis does not include restoration of function, and every developed focus of disease becomes a permanent blind spot in the field of vision. The inner layers of the retina may or may not atrophy. Should they be invaded by the disease, secondary optic atrophy results and the disc presents the characteristic yellowish tinge of retinitic atrophy (Stergm. 22).

**DIAGNOSIS.** The ophthalmoscopic picture (Stergm. 20) is typical of choroiditis disseminata and is unlikely to be mistaken for any other condition. When choroiditis disseminata occurs in a myopic eye it is apt to be overlooked and the lesions attributed to myopia. In such cases, the diagnosis of choroiditis would rest upon the wide distribution of the lesions, excessive proliferation of pigment and eruptive character of the disease. Coloboma of the macula, senile choroidal atrophy, posthemorrhagic and posttraumatic cicatrices form single or local lesions while the cicatrices of choroiditis disseminata are distributed throughout the fundus.

**STEREOGRAM 21. CENTRAL CHOROIDITIS, COMMENCING CICATRIZATION.** Left eyeground of a man, 62 years of age. Thirty-five years ago had a chancre and cutaneous eruption. Wassermann reaction for syphilis, positive. Two years ago had persistent headache and vomiting which disappeared under antisyphilitic treatment. Six months ago vision became "foggy," and since then it has steadily deteriorated. Present vision 10/200 in each eye. Sees best in a dim light, scarcely at all in sunlight (nyctalopia). Small central scotomata and concentric contraction of the visual fields. Argyl-Robertson pupillary reaction. The pathologic changes in each eye are symmetrical. The central temporal quadrant of the fundus is riddled with small, closely aggregated lesions of exudative choroiditis. In the center of the diseased area are many white spots encircled by pigment, which represent granulomata that have been converted into scar tissue. Evidently, the disease is spreading peripherally, as shown by groups of recently developed, pink colored, nonpigmented spots in the outer zone

of disease. Several superficial coal-black, branching figures and specks are seen in front of the spots. These are formed by pigment from the tessellated epithelium which has been carried into the inner layers of the retina. The optic nerve presents the picture of postinflammatory atrophy. The disc is white, but the lamina cribrosa is hidden, the retinal vessels are narrowed and the nerve is surrounded by a ring of atrophic choroid.

**DIAGNOSIS.** The symptoms presented by this case are complicated but the preceding course of the disease may be conjectured. Probably, it was as follows. Two years ago intracranial syphilis with vomiting, headache and choked disc which subsided under antisyphilitic treatment. Later, the constitutional dyscrasia reappeared as central choroiditis. The low central vision may be attributed to choroiditis, but the contracted fields and nyctalopia probably depend upon disease of the nerve and inner retinal layers. In the absence of all history the ophthalmoscopic diagnosis would lie between central choroiditis and senile degeneration with colloid excrescences. The lesions of the two conditions, however, differ as follows:

In old central choroiditis the lesions are deeply situated, vary in shape and size and tend to coalesce; colloid excrescences are very superficially situated, uniform in size and do not coalesce. In choroiditis the recent spots are pink in color and old spots are white and pigmented; colloids are uniformly of a light amber color.

**STEREOGRAM 22. CHORIORETINITIS PIGMENTOSA. RETINITIC OPTIC ATROPHY (Yellow Atrophy).** Left fundus oculi of a boy 17 years of age. Father contracted syphilis and infected mother about five months before the patient's birth. Six succeeding children were all born dead. Patient presents the stigmata of congenital syphilis. Both eyes affected with chorioretinitis but in varying degrees. Cannot see well in a dim light. *Left Eye:*—Vision reduced to mere perception of light. The most conspicuous ophthalmoscopic feature is profuse pigmentation of the retina. The pigment is situated in the inner layers, as shown by its branching, coral-like forms and tendency to cover the retinal vessels. The pigment figures are distributed throughout all regions of the retina but are most numerous at the equator where they form a continuous black meshwork. Pigment emigration, in this fundus, was caused by diffuse disease of the inner layer of the choroid. The outer layer of large choroidal vessels does not appear to be diffusely affected. In certain places, however, all layers of the choroid are destroyed and replaced by white cicatrices. The two

large scars are composed of cicatricial tissue which sends prolongations out over the surface of the retina. The small, round black spot in the lower part of the picture may be interpreted as a cicatrix in which the retinal epithelium has succeeded in completely covering the defect. The optic nerve has the dirty, yellowish-white color which characterizes post retinitic atrophy. The retinal vessels are uniformly reduced to mere threads. On and near the disc the vessel walls are white. The lamina cribrosa does not appear and the entire disc appears obscured as by a thin veil.

The layer of pigmented, tessellated epithelium which lines the inner surface of the choroid is known as retinal epithelium because it develops from the same blastodermic layer as the retina. Anatomically and pathologically, however, it is a part of the choroid and participates in its diseases. That the epithelial layer may be a primary seat of disease is extremely doubtful, inasmuch as it contains no blood vessels and is dependent upon the choroid for nutrition. The individual epithelial cells appear to possess migratory powers and when deprived of nutrition, through choroidal disease, they may wander out into the retina. These wandering cells either lose their pigment and perish or form attachments and proliferate (Fig. 37). In diffuse choroiditis not attended by profuse exudation, pigment discharged from the retinal epithelium is transported by leucocytes or lymph currents and deposited around the arterioles and capillaries of the retina, where it forms branching lines and figures the ophthalmoscopic appearance of which is aptly compared to bone corpuscles or Haversian canal systems.

This fundus exhibits a few cicatrices of deep, focal choroiditis, and also universal disturbance of the retinal epithelium indicating diffuse disease of the choriocapillaris. Choroidal disease does not usually affect the inner layers of the retina, but in this eye these layers are highly atrophic as manifested by pigmentation, extremely narrow blood vessels and yellow atrophy of the nerve. As the nutrition of the inner layers is independent of the choroid, their destruction depends on either inflammation of both choroid and retina due to action of a common toxin or, what is more probable, inanition of the retinal tissue due to blocking of the lymph channels with pigment.

**DIAGNOSIS.** This affection is frequently mistaken for retinitis pigmentosa. In this particular case the presence of choroidal cicatrices would prevent such an error. Frequently, however, the resemblance is so close



that differentiation is extremely difficult. The following differential points are applicable to the majority of doubtful cases. The blood reactions for syphilis should also be employed.

#### CHORIORETINITIS PIGMENTOSA.

May commence at any age.  
 Not a family disease.  
 Night blindness appears in advanced cases.  
 May commence in any part of the fundus.  
 May attack but one eye.  
 Development and progress may vary in the two eyes.  
 Central vision usually affected early.  
 Diligent search usually reveals white scars in fundus.  
 Irregular distribution of pigment  
 Only physical defects are stigmata in cases due to syphilis.  
 Disease may be arrested.  
 If central or scattered scotomata can be demonstrated it is not a case of retinitis pigmentosa.

#### RETINITIS PIGMENTOSA (Stergm. 53).

Commences in infancy or early childhood.  
 Reappears in a family for generations.  
 Night blindness invariably present from onset of disease and precedes pigmentation.  
 Commences anterior to equator and advances slowly backward.  
 Reaches macula in adult life (30-50).  
 Invariably, affects both eyes.  
 Develops simultaneously and progresses with great uniformity in both eyes.  
 Central vision usually retained until last stage of disease.  
 Choroidal cicatrices never present in uncomplicated cases.  
 Pigment uniformly distributed.  
 Large percentage of cases associated with body malformations, idiocy, or deafmutism.  
 Progress of disease never arrested.  
 The typical visual field is concentric contraction which commences as ring scotoma.

STEREOGRAM 23. TUBERCULOSIS OF THE CHOROID. Right eyeground of a man 21 years of age. A sister has pulmonary tuberculosis. Left eye normal. In the right eye disturbance of vision commenced two months before coming under observation—at which time it was reduced to hand movements. Vitreous was cloudy and filled with floating opacities. With

the ophthalmoscope the fundus appeared dark, except a large area on the temporal side which gave a white reflex like a bright light seen through dense fog. No fundus details discernible. Skin test for tuberculosis positive. Under full injections of tuberculin there was sudden aggravation of the fundus conditions accompanied by intense headache, hacking cough and fever. The increased eye disturbance was regarded as a local reaction to tuberculin and positive evidence that tubercular choroiditis was present. After subsidence of the reaction, tuberculin was resumed in lessened doses. Improvement followed, terminating in recovery with central vision of 20/30 but with an absolute scotoma corresponding to the choroidal lesion.\*

The stereogram was made four months after treatment was begun. The vitreous opacities have disappeared. The disc is reddened. Retinal vessels normal. Just beyond and slightly above the macula is a round, yellowish-white patch with a diameter about three times that of the disc. It is surrounded by a number of similar but much smaller spots. All other parts of the fundus are free from disease. The lesions described are supposed to be a herd of tubercles. The large patch has a well defined, soft outline. The retinal vessels which curve over its surface are elevated 0.5 mm. above the normal level of the fundus. The surface of the patch is entirely depigmented but along its inner border pigment has emigrated and settled around the descending macular vein. Beyond the outer border of the patch the retinal pigment is thinned, exposing the choroidal vessels. The smaller deposits resemble the large one.

Tuberculosis has long been regarded as a probable cause of obscure cases of choroiditis, but only since the employment of tuberculin in diagnosis has it been demonstrated that the proportionate number of tuberculous cases is large. Formerly, but two types of tuberculosis were recognized in the choroid, *i. e.*, miliary tubercle which appears in the late stage of general miliary tuberculosis, and conglomerate tubercle which forms an intraocular tumor. At the present time it is believed that tuberculosis may appear also as an exudative choroiditis, focal or diffuse, and that the ophthalmoscopic picture is by no means constant. Pure tubercle is a non-inflammatory, nonvascular growth which replaces the tissue in which it grows; facts to be borne in mind when diagnosing this disease. Authors have remarked the absence of pigmentation in early tubercular lesions of the fundus. Conclusive observations upon this point are lacking, but it

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\* Patient of Dr. Wm. G. Reynolds.

is probable that the chromophores of the choroid and retinal epithelium are completely destroyed and replaced by tubercle. When, however, a tubercular lesion of the choroid heals, the surrounding epithelium proliferates and endeavors to repair the damage as in loss of tissue from other causes, consequently, a posttubercular cicatrix is pigmented. The amount of disturbance created in adjacent tissues by intraocular tubercle appears to vary according to the location of the deposit. In the ciliary body I have seen tubercle perforate the sclera with little or no intraocular disturbance (Fig. 225). In the choroid, tubercle usually produces edema or flat detachment of the retina and cloudiness of the vitreous. Nevertheless, from the known nature of tubercle, it is probable that the disturbance is mechanical rather than inflammatory. All tissues crumble before the advance of tubercle; thus, destruction of the lamina vitrea permits fluid to pass freely through the retina into the vitreous (Fig. 53). The retina may be slightly elevated by fluid, but usually is invaded before detachment occurs. When tubercle invades and closes the retinal vessels, hemorrhages may ensue. Axenfeld and Stock hold that there may be tuberculous disease of intraocular blood vessels without visible fundus changes. Miliary tubercles appear to create little local disturbance. Quiescent tubercle of the choroid is prone to relapse; thus, an old cicatrix, in which tubercle bacilli are enclosed, may break down and become active. It is only tubercular scars that act thus as a nidus of disease in the choroid. Tuberculosis affects the choroid in connection with remote parts, although it does not appear possible for an intraocular deposit to be the only tubercular focus in the body. Certain forms of tuberculosis have a tendency to deposit metastases in the choroid, *i. e.*, tubercular meningitis of children and general miliary tuberculosis. On the other hand, the choroid is seldom affected in pulmonary tuberculosis. Miliary tubercles appear in the choroid as very small, softly outlined spots, at first grey but later yellowish in color. They rapidly increase in size but do not exceed  $\frac{1}{3}$  disc diameter (Colored Plate 2). The presence of miliary tubercles in the choroid is of great importance in diagnosing doubtful cases of continued fever.

Pathologists recognize three forms in which tuberculosis occurs in the choroid: (1) Miliary tubercle, present in nearly all cases of general, miliary tuberculosis. Miliary tubercles are most frequent around the disc but are found in all parts of the choroid. They are most frequently found in the outer layers where they project outwards into the suprachoroidea, but they may be deposited in all layers. As a rule, they are discrete although fusion may occur. The developed nodules exhibit the structure

**COLORED PLATE II**



**Tuberculosis of the Choroid**







Fig. 53.

Tuberculosis of the Choroid.

Tubercle, shown at T in the section. The retina over it is elevated by fluid.

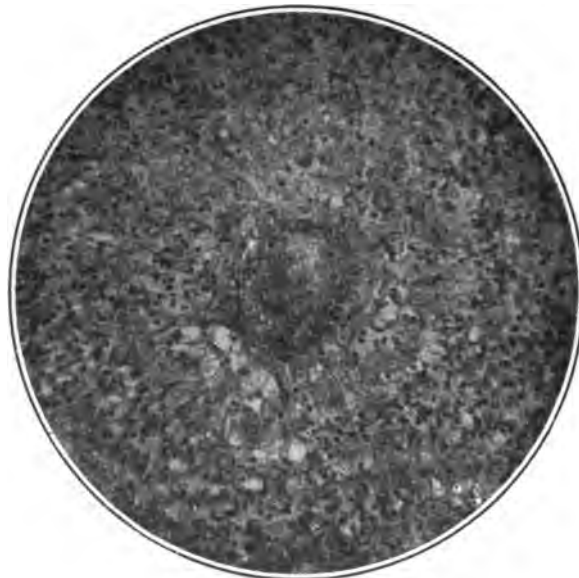


Fig. 54.

Typical Miliary Tubercle.

(Specimen from the Iris.)

typical of miliary tubercle, consisting of the Langhans type of giant cell surrounded by epithelioid and mononuclear cells (Fig. 54). Tubercle bacilli may or may not be demonstrable in the tissues. (2) Diffuse uveal tuberculosis. The entire uvea is involved. The choroid may be uniformly thickened by development of granulation tissue, containing giant cells and poorly developed tubercle systems. The advancing edge of the diffuse type exhibits tubercular nodules. The disease may terminate by perforation of the sclera or the granulation tissue may organize into scar tissue as occurs in exudative choroiditis. (3) Solitary or conglomerate tubercle which occurs in any part of the uveal tract. When fully developed it forms an intraocular tumor which may closely resemble glioma retinae. There is no doubt that single tubercles of large size develop in the choroid and subsequently undergo cicatrization.

**DIAGNOSIS. MILIARY TUBERCULOSIS.** The best evidence that recent, choroiditic spots are tubercular is a coexisting general miliary tuberculosis or tubercular meningitis. On the other hand, it occasionally happens that the nature of an obscure fever or cerebral disease is determined only by discovery of miliary tubercles in the choroid.\* As distinguished from choroiditis disseminata, miliary tubercles appear in the choroid as small, points which grow very rapidly until they attain a maximum size of about  $\frac{1}{3}$  disc diameter. The spots of choroiditis, on the contrary, change very slowly. Miliary tubercles are nonpigmented, while choroiditis is attended by more or less pigmentation. The constitutional disturbance of miliary tuberculosis is absent in choroiditis.

**CONGLOMERATE TUBERCLE (tuberculoma)** of the choroid occurs almost exclusively in the young. Occurring in those under five years of age it requires to be differentiated from glioma of the retina. Both conditions are rare. Positive local reaction to an injection of tuberculin would be conclusive evidence that the tumor was tubercular. Cases are reported of tuberculoma with extensive detachment of the retina, ophthalmoscopically indistinguishable from glioma. Glioma is said to cast no shadow in transillumination of the eye. Theoretically, tuberculoma of the choroid should cast some shadow in transillumination since it grows in intimate contact with the sclera, which is not the case in glioma. When conglomerate tubercle of the choroid is ophthalmoscopically visible, it appears as a light colored tumor accompanied by neighboring, smaller nodules. The presence of these "daughter" deposits is regarded as

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\* Tuberculin usually fails as a means of diagnosis in the late stages of general tubercular infection.



strong evidence that a tumor is tubercular. Tubercle is a nonvascular growth, consequently, if adventitious blood vessels are seen on the surface of a tumor, they point to glioma or sarcoma.

SEPTIC RETINITIS of Roth is a punctate condition of the fundus which may appear in the late stage of sepsis. The spots are whiter than tubercles and usually accompanied by hemorrhages. In the absence of history, however, differentiation from miliary tuberculosis would be extremely difficult.

FOCAL TUBERCULOSIS OF THE CHOROID. Cases of tubercular patches in the choroid which have been studied with the ophthalmoscope are so few in number that a characteristic fundus picture of the condition has not, as yet, been formulated. Nevertheless this is the most common type of intraocular tuberculosis. It appears in all parts of the uveal tract. Choroiditis in children and young adults is, in the absence of syphilis, presumably tuberculous. The best evidence that a choroiditis is tubercular is *local* activity in the diseased area following injections of tuberculin. In all doubtful cases the blood reactions for syphilis should also be employed.

The following table will indicate the chief points of distinction:

#### FOCAL TUBERCULAR CHOROIDITIS.

Frequently unilateral usually confined to a limited area.  
(May spread throughout fundus).  
Extends by development of "daughter" tubercles in proximity to original tubercle.  
Old cicatrices may be starting point of a fresh outbreak.  
Surface usually more elevated than in exudative choroiditis.

#### EXUDATIVE CHOROIDITIS.

Usually bilateral.  
Widely disseminated.  
Appears simultaneously throughout wide areas.  
Cicatrices once formed remain quiet.  
Surface not elevated except in cases of subretinal effusions and edema.

STEREOGRAM 24. DIFFUSE CHORIORETINITIS, ATROPHIC STAGE. RETINITIC OPTIC ATROPHY (Yellow Atrophy). Left eyeground of a man 41 years of age. No history or symptoms of syphilis. Wassermann reaction negative. Several immediate relatives have been affected with tuberculosis. The right eye has normal vision and the fundus presents no evidence of disease. In early manhood painless loss of vision occurred in the left eye. Central vision is 20/100. The visual field contains

large irregular defects which correspond to areas of destruction in the choroid. The ophthalmoscopic picture is that of wide-spread vascular degeneration of the choroid, and moderate atrophy of the retina. Except in the macular region, the retinal epithelium has lost its pigment, thereby exposing the vessels of the choroid. A few masses of this pigment are scattered throughout the retina. The disc is surrounded by a broad, white area in which the choroid has undergone extreme atrophy.

It is noticeable that in nearly all cases of wide-spread atrophy of the choroid the process is furthest advanced around the nerve. The reason for this is not altogether clear, although it may be due to the close adhesion between choroid and sclera that normally exists in this region, while elsewhere the two membranes are separated by lymph spaces. The retina is somewhat atrophic as shown by pigmentation, slight narrowing of the retinal vessels and, especially, by yellow atrophy of the nerve. On the outer side of the macula is a large atrophic patch in which only a few of the larger choroidal vessels remain. Similar patches exist in the periphery of the fundus which do not appear in the picture. A sclerosed choroidal vessel may appear like a solid, white band, nevertheless, it may contain a lumen through which blood circulates. This choroid has passed through a diffuse inflammation of sufficient intensity to cause irregular sclerosis of the vessels. In those regions of the fundus presenting a red reflex the retinal epithelium has not been entirely dispersed, and the dark intervascular spaces of the choroid indicate the presence of choroidal pigment. The dirty-yellowish patches around the disc and elsewhere, mark the site of an intense inflammatory process which has destroyed the pigmented cells and vessels of the choroid. After inflammation of the choroid has subsided the membrane may pass through the varied stages of tissue degeneration including the deposit of calcareous matter and formation of bone. These advanced degenerative processes usually occur in soft eyes although bone may form in a sclerosed area around the disc even though eye tension is not lowered.

The cause of choroiditis in this eye was not ascertained. Whatever the toxin, it was no longer active when the patient came under observation. Syphilitic choroiditis usually, but not invariably, affects both eyes. Ocular tuberculosis, on the other hand, generally is unilateral and it is not improbable that this is a picture of healed tuberculous lesions. I once saw a similar eyeground in a young woman, in which tuberculosis developed in an

old cicatricial patch. This form of choroiditis differs from choroiditis disseminata in being unilateral and non-progressive, also, by its patchy, irregular distribution.

STEREOGRAM 25. VASCULAR DEGENERATION OF THE CHOROID (Sclerosis of the Choroid). ATROPHY OF RETINA AND NERVE. Right eyeground of man 32 years of age. Six years ago a violent blow on the right brow produced complete and permanent blindness. It is probable that an orbital fracture occurred which extended through the optic canal and lacerated the optic nerve. The same injury, in some way, closed the ophthalmic artery, thereby interrupting the blood supply to the choroid and retina. The ophthalmoscopic picture is the same as follows optico-ciliary neurectomy. The vessels of the choroid are highly sclerosed but not entirely bloodless. Degeneration is furthest advanced around the disc where the vessels are converted into white strands of connective tissue. At the equator the fundus presents a red reflex. The retinal epithelium has parted with much of its pigment some of which has emigrated and formed branching, spider-shaped figures in the retina. The inner layers of the retina are known to be highly atrophic by the extreme narrowing of the main vessels and disappearance of all small branches. The optic nerve is very atrophic, sharply outlined and slightly excavated. It is unusual to see complete excavation of the disc in a nonglaucomatous eye, but in this case it may legitimately be due to extreme atrophy and contraction of the nerve. The same condition is occasionally found in microscopic examinations of atrophic bulbs in which the retina is not detached (Fig. 55). In the eye shown in the stereogram the tension is normal, the media clear and there is no evidence of iritis or other inflammation; therefore, the fundus changes may be attributed to malnutritive degeneration of the choroid and retina. This interpretation of the ophthalmoscopic picture is based on the results of animal experimentation. Thus, Wagenmann cut the ciliary arteries which supply the choroid without injuring the retinal vessels. The result was degeneration of the choroid, destruction of the retinal epithelium, pigmentation of the retina and degeneration of its outer layers. On the other hand, section of only the optic nerve and retinal vessels without disturbing the ciliary arteries, resulted in degeneration of the inner retinal layers but there was no degeneration of the choroid or pigmentation of the retina. From these experiments it is assumed in this case that the blood supplies of both choroid and retina were interrupted. If this explanation be accepted, why does blood continue to circulate in the vessels? It is a clinical fact, which I have witnessed, that after a tumor

of the optic nerve has been excised, and all vessels which enter the eye posteriorly cut, a meager circulation reappears in the retina within a few hours. In such cases it is assumed that the blood enters the choroid through the recurrent branches of the anterior ciliary arteries and reaches the retina through the collaterals around the head of the nerve (circle of Zinn).\*

**DIAGNOSIS.** In this fundus sclerosis of the choroid is so prominent that the condition of the nerve and retina might easily be overlooked. That the origin of the degenerations is traumatic and involves the optic nerve is suggested by the absolute blindness of the eye and extreme atrophy of the disc. The condition is differentiated from senile degeneration of the choroid by its wide extent and limitation to one eye. It differs from exudative choroiditis by its uniform distribution and absence of deep choroidal cicatrices. From diffuse choroiditis the differentiation is more difficult, but in this affection the lesions are distributed with considerable irregularity and the atrophy which follows is less uniform. The condition could scarcely be mistaken for albinism. Moreover, in none of the diseases mentioned is the eye completely blind.

**STEREOGRAM 26. ATROPHY OF CHOROID. ACQUIRED ALBINISM.** (Depigmentation of Fundus in a Diabetic). Right eyeground of a woman 53 years of age. Mother of eight healthy children. Although well nourished and apparently healthy, the urine contains 9 per cent. of sugar. Always had good vision until two years ago, when it began to fail. Right eye counts fingers at three feet. Left eye, vision is 20/70. Visual fields contain large irregular defects. There is neither photophobia nor torpor retinæ. Has never experienced any subjective symptoms of choroiditis, *i. e.*, photopsiæ, micropsiæ, etc. Similar fundus changes exist in both eyes, but are furthest advanced in the right. In this eye there is almost complete depigmentation of the entire fundus. The choroidal vessels appear with great distinctness against the uncovered sclera, producing an ophthalmoscopic picture closely resembling that of albinism (Stergm. 15). The smaller vessels of the choroid and choriocapillaris have disappeared from large areas of the fundus which now present a white reflex. Evidently, these choroidal vessels have been obliterated by a simple atrophic process, inasmuch as no evidence of inflammatory action exists in the eye.

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\* In tumors of the optic nerve adventitious vessels form before operation, which replace the central artery and supply the retina.

The retinal vessels appear normal.\* The disc shows no sign of disease, although it appears reddened and ill defined as in all cases where it is no longer contrasted with the normal choroid. The choroidal atrophy is most marked in the posterior polar zone and, in a general way, diminishes toward the periphery. The disc is surrounded by a wide, irregular-shaped ring of highly atrophic choroid. The choroidal and retinal pigment have vanished without migrating into the retina. On the temporal side of the picture are three small groups of pigment specks, situated among the vessels of the choroid. The macular region contains a few reddish spots which have partially resisted the degenerative process. One of these contains the fovea which appears as a small, round, red spot. That this is the patient's point of fixation is proved by directing her to look at the center of the ophthalmoscopic mirror, when this red spot will be found in the observer's direct line of vision.

The iris of the congenital albino contains no pigment, consequently, an excess of light enters the eye, causing intense photophobia. In this patient the irides were normally pigmented and she had no intolerance of light. These facts tend to confirm the theory that it is the pigment in the iris, not that in the fundus, which prevents dazzling in the normal eye.

The pathologic changes in this eye may be attributed to the existing diabetes, a disease known to cause degeneration of the ocular vessels. When diabetes affects the vessels of the retina, the fundus presents the picture of retinitis. It may be asked, why degeneration of the retinal vessels should be attended by edema, exudation and hemorrhage, while degeneration of the choroidal vessels produces none of these phenomena. The explanation is found in the following anatomic variations. The retinal vessels have no anastomoses; hence, obstructive disease is followed by sudden disaster. The choroid, on the contrary, is a vascular plexus whose free anastomoses are a protection against local ischemia, edema or hemorrhage in case a vessel is obstructed. Although the vessels of this choroid are being obliterated by angiosclerosis, their walls remain invisible, differing in this respect from the familiar forms of choroidal sclerosis in which the walls of the vessels become very white. Both Raehlmann and Coats regard translucent thickening of a vessel's walls as due to disease of the inner coat (intima), and opaque thickening of the walls to changes in

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\* It was observed as a curious physical phenomenon, that the arteries exhibited central light streaks only where they crossed the nerve or in regions of the fundus which furnished a reddish background.

the external coat (adventitia). Opaque vessels are also rare in diabetic retinitis although profound vascular disease is present as shown by the number of hemorrhages (Stergm. 38). The depigmentation of this fundus may be analogous to the discharge of pigment from the iris and ciliary body that follows iridectomy in a diabetic subject. In such cases the amount of pigment discharged may cause the aqueous to look like ink.

**DIAGNOSIS.** This rare fundus condition is presented for comparison with other forms of choroidal atrophy, from which it is differentiated chiefly by translucent atrophy of the vessels and destruction of pigment. It may be distinguished from congenital albinism by recent failure of vision, normal pigmentation of other parts and atrophy of the choroidal vessels. The eye of the albino is normal except that it contains no pigment.

**STEREOGRAM 27. SENILE CHANGES IN THE FUNDUS. COLLOID EXCRESCENCES (Drusen). MACULAR DEGENERATION. ARTERIOSCLEROSIS.** Left eyeground of a woman 73 years of age. Similar fundus conditions present in both eyes. Vision 20/50. The region about the nerve and macula is covered with superficially situated, round, yellowish dots, none of which exceeds in size the width of the central vein. They are confined to the posterior polar zone and occur chiefly on the temporal side of the nerve. The dots occur singly and in groups. Some of them are closely crowded together, but show no tendency to coalesce and form large masses. They are always below the retinal vessels, but some of the larger dots appear elevated above the surface of the retina. Neither the dots nor surrounding fundus are pigmented. At the macula the foveal pit is surrounded by a ring of fine, granular pigment. The optic nerve contains a deep physiologic excavation with a very white floor, on which markings of the lamina cribrosa are very distinct. These appearances are within physiologic limits and the nerve may be considered normal. Arteriosclerosis of the retinal vessels is indicated by undue brightness of the central light streak on all arteries and, particularly, by the white walls seen on an ascending artery as it crosses a vein. All the morbid changes observed in this eye are degenerative in character and incident to old age.

The yellow dots, so conspicuous in this fundus, are known under the various names of colloid, hyaline, and warty excrescences, guttate chorioiditis, colloid degeneration of the choroid, etc. Recent English writers have adopted the German name "drusen" to designate these bodies. A few colloid bodies are ophthalmoscopically visible in the eyes of most very old people. Occasionally they appear in great numbers, principally at the posterior pole. Similar bodies are seen in all forms of pigmentary

degeneration of the choroid and they are supposed to constitute the spots found in retinitis punctata albescens. These dots do not themselves injure vision, but frequently are associated with senile degeneration of the retina or choroid in which case vision may be very low.

In the eyes of people over 50 years of age and in eyes which have undergone degenerative changes, the microscope nearly always reveals the presence of small, globular or hemispheric elevations, situated on the inner surface\* of the lamina vitrea which, from their appearance, have been known as colloid or hyalin bodies (Figs. 56-57). On their inner surface, these bodies are completely or partly covered by the pigmented retinal epithelium to which they are intimately connected. They may appear to form a part of the lamina or be connected only by a narrow neck. Sometimes they are found free in the retina. Loosely attached and free drusen are supposed to have separated themselves from the lamina, but there is no evidence that such bodies were ever connected with it. Colloid bodies may lose their covering of pigmented epithelium in which case they become ophthalmoscopically visible. On the other hand, an eye may contain a great number of colloids, but if covered with pigment they cannot be seen with the ophthalmoscope.

The origin and nature of these formations have not been definitely determined. In structure they resemble the lamina vitrea and, by some, are regarded as a proliferative thickening of that membrane. Others consider them as degenerated epithelial cells. The lamina vitrea is a homogeneous membrane deposited by the pigmented epithelium and it is improbable that it possesses proliferative power. It is more reasonable to suppose that the excrescences on its surface originated from the same source as the membrane, *i. e.*, that they are a deposit from the cells of the pigmented epithelium. In diseased eyes the colloids are deposited by the cells in response to an irritant.

The pigment which surrounds the fovea in this eye marks the early stage of senile, chorioretinal atrophy. The macula of the aged frequently exhibits pigment and irregular, yellowish spots. These changes depend on vascular degeneration of the choriocapillaris, whereby the retina is deprived of nutriment. The resulting degeneration is visible chiefly at the macula, for the reason that in this region the retina consists only of outer layers. Senile macular degeneration may result in complete loss of

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\* Excrescences never form on the outer surface of the lamina, a fact which suggests their origin from the retinal epithelium.

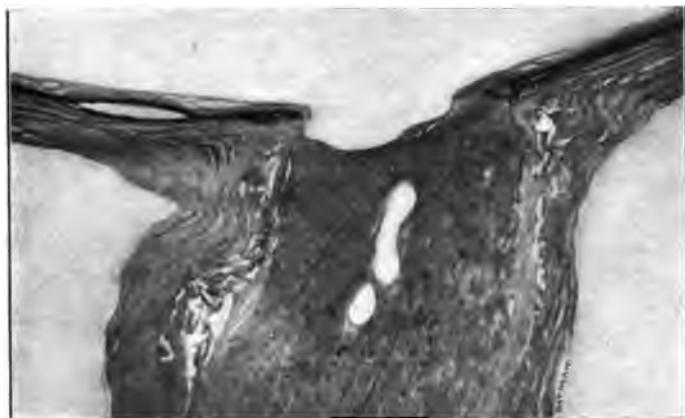


Fig. 55.

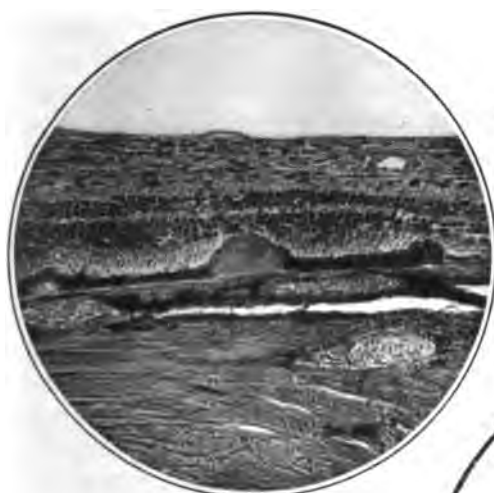


Fig. 56.

Fig. 56-57—Colloid Degeneration of the Choroid. (Drusen.)

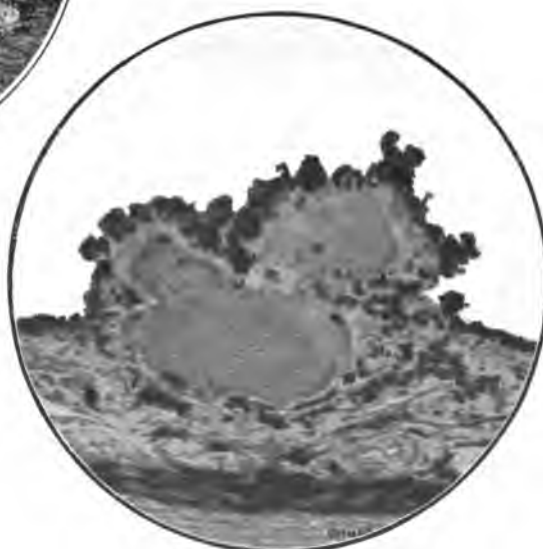


Fig. 57.

Fig. 55—Complete Non-glaucomatous Excavation of Disc. Due to Extreme Atrophy of the Optic Nerve. Specimen from an Atrophic Bulb.





vision. When senile atrophy of the choroid extends to the large vessels, white, sclerotic patches develop, especially around the disc and macula. The deterioration of vision in the case presented above was due to the macular changes, not to the colloid excrescences.

Retinal degeneration is a constant feature of old age. The membrana limitans and the fibers of Müller are greatly thickened and the nervous elements may degenerate. In consequence of these changes the retina becomes less transparent and gives a stronger ophthalmoscopic reflex. The most constant senile retinal degeneration occurs in the periphery. Here the retina is thickened and divided by Müller's fibers into cyst-like spaces which contain no trace of nervous elements (Fig. 58). From its situation and from the appearance of the spaces this form has been termed peripheric cystic degeneration of the retina, or Iwanoff's edema.

Arteriosclerosis of the retinal vessels is nearly constant in advanced age. The changes are not always ophthalmoscopically visible. Senile sclerosis of the retinal vessels has not the same evil prognostic import as arteriosclerosis from causes other than age.

Senile alterations in the optic nerve are secondary to either retinal degeneration or to arteriosclerosis.

**DIAGNOSIS.** A punctate condition of the fundus, limited to the posterior pole, occurring in an aged person is, presumably, due to colloid excrescences. Other punctate conditions which might be mistaken for senile colloids, differ from them as follows. Gunn's dots (Stergm. 16) occur in the young, are smaller in size and far less distinct than colloid bodies. Retinitis punctata albescens (Stergm. 54) commences in early life, is slowly progressive and associated with night blindness. The dots appear first at the periphery of the fundus. The lesions of central choroiditis (Stergm. 19) are differentiated by their deep situation, large size, irregular shape, presence of pigment and tendency to coalesce. Furthermore, some of the spots are, obviously exudative in character.

**STEREOGRAM 28. RUPTURE OF THE CHOROID.** Left eyeground of boy 10 years of age. One year ago the eye was struck by a missile from a sling. At that time the fundus was obscured by blood in the vitreous. Three months later sufficient blood had been absorbed to disclose a large white patch around the nerve but details were not discernible. The stereogram shows the condition of the fundus twelve months after the injury. The blood has been completely absorbed from the vitreous, leaving the media clear. An unusually extensive rupture of the choroid is seen completely encircling the nerve. Probably, the laceration commenced

between the disc and macula, but was prolonged far beyond the usual length. Observe the tendency for the rupture to approach the nerve. The course of the rupture is concentric with the outline of the disc. In shape it is wide in the center and narrower at the ends. In the supranasal region is a smaller rent in the choroid with a lateral extension and forked extremity. The retina is not torn, as shown by its vessels which pass uninterruptedly across the ruptures. For some distance beyond the rupture, the retina is darkened by fine, dust-like pigment, signifying that the pigmented epithelium was disturbed beyond the border of the fracture. Along the edge and over the surface of the exposed sclera are several groups of pigment. The pigment along the edge is proliferating retinal epithelium which is endeavoring to cover the hiatus. This pigment will progressively increase in amount.

Rupture of the choroid usually is produced indirectly by sudden, violent compression of the eyeball. In most cases the force is applied to the front of the eye by a blunt body, like a fist, a missile, cork from a gas charged bottle, a ball, etc. Exceptionally, it is caused by force which fractures the skull. The location and form of the rupture are characteristic. Almost invariably it occurs at the posterior pole, within two or three disc diameters of the nerve (Colored Plate 3). Statistics indicate that it is about five times more frequent on the temporal than on the nasal side. Ruptures confined to the region above or below the nerve are uncommon. The favorite site is between the macula and disc. Immediately following the accident there is hemorrhage which usually enters the vitreous and hides the fundus. Vision, of course, is interrupted. After resorption of the blood, rupture of the choroid is recognized by discovery of a white, crescentic figure at the posterior pole, the long diameter of which is concentric with the margin of the disc. The borders of the rupture are generally pigmented. Blood may remain in the rent for a long time. The rupture may be either single or multiple. Frequently the rent is forked or branched. The tear involves the entire thickness of the choroid and layer of pigmented epithelium. The suprachoroidea also is torn, but shreds of this membrane may remain in the wound, attached to the sclera. Yellowish streaks, in form and position resembling ruptures, may follow trauma or accompany lacerations of the choroid. These streaks are supposed to be incomplete fractures of the choroid and pigmented epithelium, which have not extended through to the sclera. Rupture of the choroid frequently is complicated with dislocation of the lens, detachment of the retina, rupture of the sphincter pupillæ, etc. The group of cases

**COLORED PLATE III**



**Rupture of the Choroid**  
**(Situated on the Nasal Side of the Disc)**





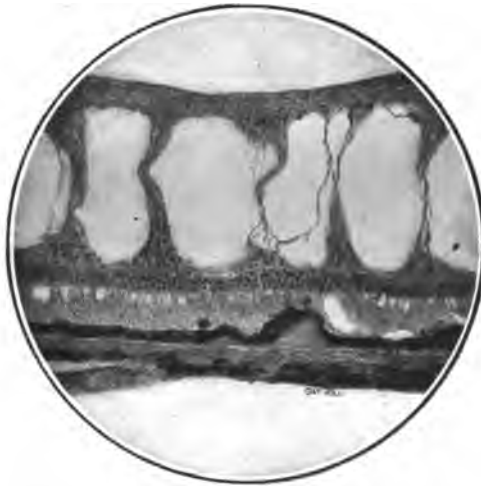


Fig. 58.

Fig. 58—Senile Degeneration of the Retina.

Cyst-like spaces devoid of nervous elements and separated by Müller's fibers have formed in the thickened retina.

Fig. 59—Rupture of Choroid, Extending into Nerve Sheath. N, optic nerve; D, detached retina; H, hernia of retina into ruptured sheath.



Fig. 59.



Fig. 60.

Fig. 60—Rupture of Choroid extending into Optic Nerve.

described in this chapter includes only those ruptures of the choroid in which injury is repaired without loss of the eye. In a larger number of nonperforating injuries, laceration of the choroid is so extensive and the resulting hemorrhage so profuse that the eye is irrevocably lost and enucleation follows, without the choroidal lesion ever coming under observation. In a number of such cases, microscopically examined, I have noted a tendency for extensive laceration of the choroid to extend to the edge of the nerve or even into its substance. A curious example is shown in Fig. 59. The photomicrograph is from an eye which, thirteen months before enucleation, was blinded by a blow from a stone projected by a blast. Microscopic examination shows an extensive fracture of the choroid which is prolonged through the sclerochoroidal ring into the inter-vaginal space around the nerve. Through the aperture thus created a knuckle of detached retina has prolapsed into the sheath of the nerve, forming a hernia. Figure 60 is from an eye enucleated ten days after violent application of blunt force to the ciliary region. No external rupture of the globe. Microscopic examination revealed a detached retina, large subretinal blood clot and a broad rupture of the choroid, one end of which extended into, and partly through, the head of the optic nerve.

It is difficult to understand why rupture of the choroid always occurs at the posterior pole, usually on the temporal side, and follows a circumpapillary course. The number of theories offered in explanation is a measure of their inadequacy. These are, (1) *contre coup*; (2) driving the eyeball over the nerve; (3) rotation of globe which suddenly is arrested by the optic nerve or, (4) by the short ciliary arteries. The position of the classic rupture of the choroid corresponds to the posterior border of the oblique muscles and it is probable that, when the sclera and attached choroid are forcibly driven backward, these muscles contract spasmodically, and that the choroid is broken across their rigid edge. The retina escapes laceration because it is free and unattached.



## CHAPTER V.

### SCLEROSIS OF THE RETINAL VESSELS (*ANGIOSCLEROSIS*) AND RETINAL HEMORRHAGE.

In the following paper an effort has been made to present the pathologic events of arteriosclerosis in sequence, although, clinically, no disease is more erratic in its course.

The first ophthalmoscopic evidences of sclerosis of the retinal vessels are dilation, tortuosity and pulsation of the arteries. Thoma considers the dilation as due to loss of elasticity and contractility in the walls. Naturally, dilation is followed by tortuosity. The pulsation is characterized by a lateral displacement of the whole artery, most marked at bends and curves. When present, it is well developed and unmistakable. It is a true arterial pulsation termed *locomotion pulse* as distinguished from the pressure pulse seen in glaucoma and whenever general blood pressure sinks below intraocular pressure. Locomotion pulse occurs in all cases of incipient retinal angiosclerosis in which the arteries are primarily affected, but as it causes no disturbance of vision it is seldom seen by the ophthalmologist.\* As vascular degeneration progresses and the arteries become rigid, arterial pulsation disappears. Observations are lacking as to the transparency of the arterial walls in the first stage of arteriosclerosis, but in the cases which I have recognized, the walls of the arteries were distinguishable by weak illumination.

STEREOGRAM 29. DIFFUSE RETINAL ARTERIOSCLEROSIS PRIMARY STAGE. Boy, 15 years of age. Sought advice because of recurring epistaxis. Incidentally, the eyes were examined when a probable cause for the bleeding was discovered in the condition of the blood vessels. The retinal arteries were slightly dilated and more tortuous than the veins. The smaller vascular twigs were very distinct. The arterial blood columns had extremely delicate, light borders, evidently, a slight opacity of the walls. The most conspicuous feature in the fundus was a strong arterial pulse, especially pronounced wherever an artery made a sharp bend. The veins were of normal size and not unduly tortuous. The optic disc was

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\* Suggests the necessity of using the ophthalmoscope in general diagnosis.

slightly reddened. Urine was found to contain albumin and hyalin casts. Heart slightly dilated but no murmurs. Blood pressure was 120 mm. Hg.

In the early stage of typical, retinal arteriosclerosis the veins are unchanged and as a rule are less tortuous and no broader than the arteries. This disease, however, is capricious and the veins may be attacked before the arteries, in which event there is venous turgescence and no pulsation in the arteries. In the case described above, the blood pressure was not elevated. Probably, it always is normal or subnormal in the stage of arterial relaxation and dilation. Later in the disease when the arteries grow rigid and the terminals unyielding, blood pressure will be increased. In very early arteriosclerosis, the relaxed blood vessels absorb much of the force imparted to the pulse wave.

Degenerative changes in the arteries always are accompanied by analogous changes in the veins, although the disease may be further advanced in one than in the other. Nearly all the retinal vessels may exhibit disease. More often, however, the pathologic changes are limited to a certain vascular circuit or even to a single vessel. Angiosclerosis is associated with a variety of chronic diseases, to some of which it bears a causative, and to others a secondary relation. In any event, the discovery of vascular degeneration in the retina is an omen of gravest significance.

Opportunities for the microscopic study of early arteriosclerosis are rare, consequently little is known of the histologic changes occurring in the stage of arterial relaxation and dilation. Mott considers degeneration of the elastic and muscle fibers as the primary lesion. Jores says that arteriosclerosis commences by formation of spaces in the elastica, at the period of time corresponding to arterial dilation. Ribbert also describes the first change as spacing of the intima. Thoma saw the starting point in a noninflammatory swelling of the media. Vasomotor paralysis has been advanced as a cause of the early relaxation. This appears improbable; at least, section of the cervical sympathetic in animals does not affect the retinal vessels.

**ETIOLOGY.** No age is exempt from arteriosclerosis, but in infancy it is rare, while as a manifestation of senility it is constant. It is much more prevalent among young adults of both sexes than is generally supposed. It is common among muscular, hard-working individuals and comparatively rare among the feeble. Vascular disease is more common among men than women, but not where women perform the same manual labor as men and not after the climacteric. It is very frequent among those who labor in a cold, damp atmosphere. The most important factor, however, is

heredity, manifested in certain families by a tendency to vascular degeneration from slight causes and by premature senility. Arteriosclerosis is intimately associated with chronic nephritis and slight, vascular changes in the retina may antedate the appearance of albumin in the urine. Prominent causative diseases are gout, articular rheumatism and syphilis. Vascular degeneration often is traceable to the toxic action of lead, alcohol, tobacco, phosphorous and other drugs. Many recent writers regard intestinal autointoxication as one of the common causes of this disease. Infectious vasculitis may occur in any infection.

**DIAGNOSIS.** Opportunities seldom occur for diagnosing arteriosclerosis during the stage of arterial relaxation. Simple association of tortuous arteries with a locomotion pulse is almost pathognomonic of arteriosclerosis. Aortic regurgitation, aneurism and Basedow's disease may also show locomotion pulsation and these diseases must be excluded in making the diagnosis. As distinguished from the locomotion pulse, which occurs in the arteries throughout the retina, a pressure arterial pulse is confined to the disc. Pulsation in the retinal arteries should not be diagnosed unless it is distinct and unmistakable. Slight movements of the patient's eye or of the ophthalmoscope are easily mistaken for pulsation in the vessels.

As first described by Thoma, thinning and distension of the vessel walls in arteriosclerosis is soon arrested by proliferation and thickening of the intima, media and adventitia, a process regarded as compensatory. With the advent of this process the disease enters the stages of vascular rigidity and degeneration, the pathology of which includes nearly all morbid changes that occur in the inner layers of the retina. The chief ophthalmoscopic features of the degenerative stage are changes in the color of the vessels, opacities in their walls and abnormal variations in the size of the blood columns. The arteries as a whole are somewhat lighter in color than normal. In addition, the central light streak becomes brighter, broader and extends unusually far out into the periphery. When this appearance is pronounced, the vessels have been aptly compared to burnished wire and are called "silver wire arteries" (Stergm. 34). The sclerosed wall of a retinal vessel may or may not become opaque. A normal retinal vessel has transparent walls. It is the column of blood within the vessel and not the vessel itself that is seen. Therefore, an opacity, however slight, connected with a blood column should be regarded as abnormal. When the walls of a vessel become visible the blood column appears bordered by light lines, the distinctness of which

is governed by the degree of opacity. Of course, the opacity is not limited to the lateral walls, but is visible only there, because the light from the blood column renders translucent a moderate opacity in the overlying wall. Also, on the sides of the vessel we are looking through a much greater thickness of tissue than on the top. The degree of opacity in the walls of a retinal vessel is no measure of the thickening which exists. In fact, the walls may be thickened even to the point of obliterating the lumen without becoming visible. Thickening and opacity of the walls is first distinguishable and most pronounced where an artery crosses over a vein. Under normal conditions the walls of an artery do not hide an underlying vein, and the vein can be traced up to the blood column of the artery and sometimes may be faintly seen as it passes beneath. If, however, the walls of the overlying artery are opaque they obscure the vein and the course of the latter appears to be interrupted at a short distance on either side of the artery (Stergms. 30-31-32). Frequently, an underlying vein is hidden by an artery with thickened but translucent walls. In such cases an open space appears to separate the ends of the vein, which seem to be pointed, from the artery. Sometimes a sclerosed artery is visible beneath a thin normal vein. As a rule, opacities extend only for short distances along a vessel. Opacities often appear as plaques or bright spots in the walls.

Arteriosclerosis shows a preference for the large vessels on the disc, especially the central artery and vein at their point of bifurcation (Fig. 61). Arteriosclerotic changes on the disc, however, should be diagnosed with caution, inasmuch as it is a common congenital condition for the large vessels on the disc to be ensheathed with connective tissue, which appears as narrow white lines bordering the arteries and veins. As a rule, this tissue only extends to or slightly beyond the edge of the disc. As wall degeneration advances the blood column becomes irregularly narrowed. Translucent thickening of the wall, with greatly narrowed blood column, limited to short sections, is said to indicate nodular, endothelial proliferation (endarteritis nodosa).

STEREOGRAM 30. ADVANCED SCLEROSIS OF RETINAL VESSELS. Left fundus oculi of a laboring man, 59 years of age, with advanced general arteriosclerosis. Urine normal. Marked arteriosclerotic changes appear in both retinae, especially the left. The disc has a brick-red color. The arteries are narrowed but not tortuous. The walls of the primary subdivisions of the central artery are thickened but translucent as shown where they cross over a vein. At such points the underlying

vein is completely obscured for a short distance on each side of the artery, but the walls of the artery are not visible with a bright illumination. This form of thickening is supposed to indicate endothelial proliferation in the artery. In the inferior temporal vein the blood column is irregularly narrowed and, in places, bordered by white lines. These linear opacities are supposed to be in the outer walls and imply that the sclerosing process has extended to the media and adventitia. The veins are tortuous, dilated and broadened peripherally. In passing below a sclerosed artery they appear narrowed and disappear. The most conspicuous changes are found in the superior temporal vein. Between the disc and point of bifurcation, the vein is extremely tortuous and greatly narrowed except at certain places which are ectatic. Beyond the point of bifurcation the branches of the vein are abruptly and enormously dilated. A great number of small collateral veins have developed along the constricted portion. Throughout the area to which this vein is distributed, the retina is opaque and contains old and recent hemorrhages. The meshwork (*wundernetze*) of little veins consists of dilated venules which have established a collateral flow.

**DIAGNOSIS.** The ophthalmoscopic diagnosis of retinal arteriosclerosis is made only after a successful search for some of the vascular changes described above. A common error is mistaking a congenital connective tissue sheath, on a disc vessel, for a pathologic condition. The congenital tissue is limited to the disc and, although very dense, the retinal vessels are perfectly normal. Were such dense opacity due to arteriosclerosis it would be accompanied by unmistakable evidence of disease elsewhere in the retina. Healthy retinal vessels are very compressible, while sclerosed vessels are comparatively incompressible. These facts may be utilized in diagnosis. Thus, if gradually increasing pressure is applied by the finger to the eye the following phenomena may be observed with the ophthalmoscope: (1) strong venous pulsation appears on the disc; (2) arterial pulsation; (3) cessation of circulation and blanching of the vessels. Rigid vessels, on the other hand, resist pressure and, where retinal arteriosclerosis is advanced, neither venous pulsation nor blanching of the vessels can be produced by pressing on the globe with the finger. The ease with which the retinal circulation can be artificially arrested is a measure of arterial tension and mural rigidity. Melville Black states that where the heart is dilated and the blood pressure low, pressure on the globe causes decided emptying of the arteries with but little change in the veins. There are certain external ocular mani-





Fig. 61.

Sclerotic Changes in the Central Artery and Vein at their Bifurcation on the Disc.

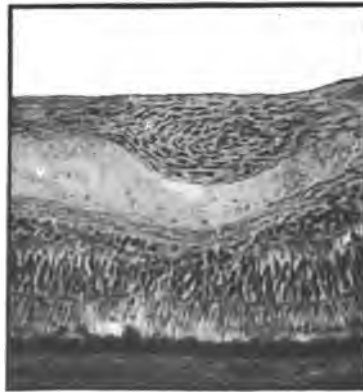


Fig. 62.

Distension of Retinal Vein, due to Pressure from the Sclerosed and Rigid Artery Overlying.

festations of arteriosclerosis of considerable diagnostic value. Spontaneous subconjunctival hemorrhage, ocular or palpebral, occurring in people over 35 years of age, usually indicates vascular degeneration. The same is true of fugitive edema of the lids. Hirschberg first noticed that persons with arteriosclerosis, especially of the cerebral vessels, were subject to recurring chemosis of the conjunctiva. This was, presumably, caused by closure of small branches from the anterior ciliary vessels. White lines, due to distension of the perivascular sheaths, border the vessels in early neuritis (Stergm. 65) and after severe concussions of the retina (Stergm. 59). Such lines are differentiated from arteriosclerotic opacity by their situation on and near the disc, by the evidently acute nature of the process and by the history. When intraocular pressure is increased by pressing on the eye with the finger, pulsation occurs in the retinal veins and arteries. The degree of pressure necessary to cause pulsation is diagnostic; thus, in normal eyes it is produced by moderately firm pressure; in glaucoma it appears when the eye is barely pressed, but it cannot be induced in a sclerosed artery by any degree of compression.

The effect of high blood pressure in the retina is felt especially in the terminal venous twigs, which become distended, tortuous and, apparently, increased in number. Retinal arteries do not retain the tortuosity acquired in the early stage of arteriosclerosis. As the arterial walls become rigid from proliferation of new tissue, tortuosity decreases in the arteries and increases in the veins. Raehlmann observed that tortuosity of the retinal arteries was most pronounced where general arteriosclerosis was but slightly developed, and entirely absent in the most highly developed cases of this disease. Arterial pulsation also disappears as rigidity develops in the arteries. Raehlmann describes a characteristic type of venous pulsation usually present in advanced arteriosclerosis. Other observers, including the writer, have been less fortunate than Raehlmann in discovering this pulse. In passing under a sclerosed artery a vein dips with a sharp twist beneath it and at the same time appears to narrow. The pointed appearance which a vein assumes as it passes beneath a semiopaque artery is an optical effect produced by the walls of the overlying artery which gradually obscure the vein. A rigid artery lying across a flaccid vein may interfere with the venous circulation, causing distension of the vein distal to the point of pressure (Fig. 62). The veins may be dilated in the peripheral parts and narrowed at the disc. This is due partly to thickening of the venous walls at the nerve and partly to accumulation of blood in the terminals and capillaries. Varicosities are fre-



quent, appearing as dark, oblong enlargements or knob-like protuberances in the course of a vein. Moderate venous distension may be the logical result of arterial rigidity, but when extreme, it probably depends upon obstructive endophlebitis in the central vein or branches (Fig. 63). Miliary aneurisms form on the small arterioles and capillaries. In the larger vessels local dilations occur where the walls are least affected. Frequently, they form immediately behind a rigid constriction. It is doubtful if pulsating retinal aneurisms occur in the retina except as the result of traumatism.

**MORBID ANATOMY.** Within the retina the arteries possess a thick wall containing elastic fibers but no elastic membrane. The veins consist only of the endothelial lining upon a thin connective tissue wall which contains no elastic fibers. Thus by employing a selective stain for elastic tissue the retinal arteries are readily differentiated from the veins. In the retina two forms of vascular degeneration predominate. In one, the intimal cells proliferate and form nodular or elongated masses on the inner wall of the vessel. Usually these deposits occur only on one side, thereby causing eccentric narrowing of the lumen. Exceptionally, nodular growths spring from opposite sides, reducing the lumen to a central slit (Artery in Fig. 64). In the other form of degeneration the walls of the vessels thicken from proliferation of connective tissue, which narrows the lumen concentrically (Fig. 65). The latter process is known as fibrosis and usually terminates in hyaloid degeneration (Vein in Fig. 64). These two forms of degeneration often are combined. An artery or a vein may be affected with either type, but the arteries tend to endothelial proliferation and the veins to fibrosis. Sometimes the perivascular sheath of a sclerosed vessel is dilated (Fig. 66). The lesion of syphilitic arteries is a dense, round celled infiltration in and around the outer walls (Fig. 67) which is followed by formation of cicatricial tissue (Fig. 68). Recently, it has been observed that organs and tissues to which arteriosclerotic vessels are distributed undergo fibrosis and hyaloid degeneration coincident with degenerative changes in the arteries. It is uncertain whether these tissue changes are wholly secondary to the vascular disease or whether both vessel and tissue changes result from the action of the same morbid influence. The deleterious effects of arteriosclerosis in the retina appear to result from impaired nutrition due to obstructions in the vessels, and its functional activity does not appear to suffer so long as circulation is not interrupted. It is possible, however, that the retinitis of albuminuria and diabetes may, in part, depend upon the local action of a toxin. Coats suggests that

**Fig. 63—Obstructive Endophlebitis.**

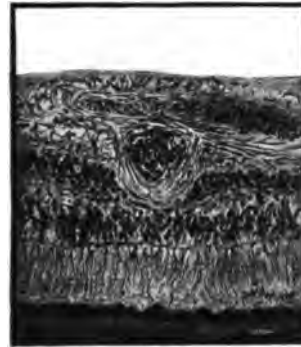


**Fig. 63.**

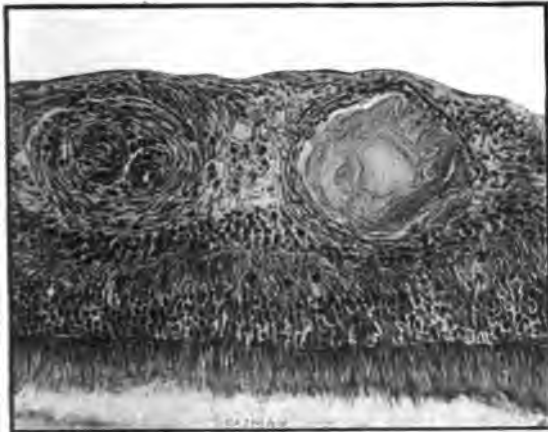


**Fig. 64.**

**Fig. 64—Nearly Complete Obstruction of Retinal Artery (shown to the left in the section) due to Nodular Growths Springing from Opposite sides of its Wall. The Wall of the Vein (shown to the right) is Undergoing Hyaloid Degeneration as the Result of Fibrosis.**

**Fig. 65—Concentric Narrowing of the Lumen from Proliferation of Connective Tissue.**

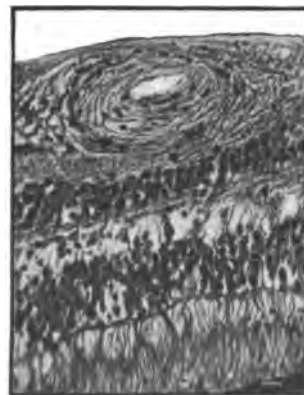


**Fig. 65.**



**Fig. 66—Dilation of the Perivascular Sheath of Sclerosed Vessel.**



**Fig. 66.**



**Fig. 67.**

**Fig. 67—Dense, Round Cell Infiltration of the Outer Vascular Walls, due to Syphilis.**

**Fig. 68—Secondary Cicatricial Formation in Vascular Walls as the result of Syphilis.**

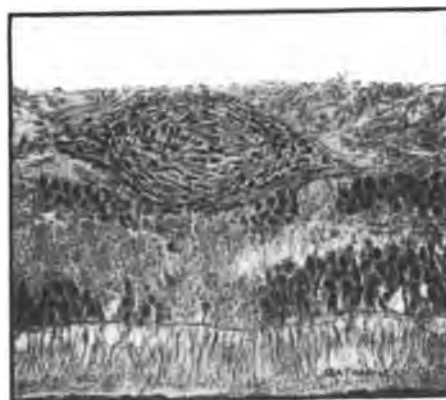


**Fig. 68.**



disease of the intima may be caused by a toxin circulating in the blood while thickening of the outer wall is compensatory to meet high blood pressure.

The new-formed tissue in the walls of a vessel may undergo retrogressive changes and pass through the various phases of fibrous, hyaloid, amyloid, fatty and calcareous degenerations. A wall affected with fatty degeneration is easily torn; consequently, hemorrhage frequently is due to this cause. In the hyaloid stage of degeneration both intima and media are replaced by a glassy, homogeneous material which causes great reduction or even obliteration of the lumen.

**STEREOGRAM 31. ARTERIOSCLEROSIS AND OPTIC NEURITIS.** Right eyeground of a man 49 years of age with general arteriosclerosis. Is of robust appearance and unusually muscular. Works in a foundry where he is accustomed to carry heavy weights. Takes several alcoholic drinks daily. Blood pressure, 260 mm. Hg. Urine contains a trace of albumin but no casts. Vision 20/20. Advanced sclerosis of the retinal vessels in each eye; most pronounced in the right. The optic nerve is very red, slightly elevated and its outlines are obliterated. The retinal arteries are pale in color, pursue a straight course and are irregularly narrowed. The veins are tortuous, dilated and their course appears to be interrupted wherever they are crossed by an artery. The arterial walls exhibit both translucent and visible sclerosis. The blood columns of the superior nasal and inferior temporal arteries are completely hidden, for short stretches, by wall opacity. The inferior nasal artery contains several slight, fusiform dilations alternating with sharp constrictions in which the blood column is reduced to a mere thread. The terminals of the veins are extremely tortuous. Varicose dilations (phlebectasiæ) exist on two veins, just below the disc. The presence of high blood pressure and rigidity of the vessel walls in this case is illustrated by external pressure on the eyeball, which fails to cause either venous pulsation or blanching of the retina, as would occur in a normal eye.

**DIAGNOSIS.** The degenerated retinal vessels in this fundus furnish the key to a correct diagnosis. In pseudoneuritis (Stergm. 13) and in congenital cyanosis (Stergm. 58) arteries as well as veins are tortuous and there is no angiosclerosis. Closure of the central retinal vein (Stergm. 46) is invariably attended by profuse hemorrhage. In papilledema of cerebral origin the nerve head presents a soft, whitish appearance and the height of the swelling is much greater than in this case. This fundus is an example of angiopathic disease, ophthalmoscopically indistinguishable from changes which might occur in Bright's disease. In fact, the conditions are identical,

and there is no doubt that vascular degeneration exists in the kidneys which, when further advanced, will cause this case to be classed as one of chronic nephritis.

### *Bibliography.*

- Adler, Med. Rec., May 10, 1902, p. 721.  
 Alleman, American Medicine, Feb. 20, 1904.  
 Black, Melville Jour. Am. Med. Assn., July 29, 1911.  
 De Schweinitz, Oph. Record, Aug., 1906. Trans. Am. Oph. Soc., 1906. Maryland Med. J., 1900. Path. Soc., Phila., 1906. Intern. Clinics, Vol. 1, Series 17, 1907.  
 Elschmig, Klin. M. f. Augk., 1898, p. 25.  
 Friedenwald, Jour. A. M. Assn. XVI, 1891; A. of O., 1896, XXV, p. 177.  
 Fuchs, A. f. A., XI, 440.  
 Gloor, A. f. A. XXXV, p. 228.  
 Gull and Sutton, Med. Clin. Trans., 1872, IV, p. 273.  
 Gunn, T. U. S., 1898.  
 Haab, WI. Versamml. d. Heidgh. Oph. Gesellsch., 1900.  
 Heller Munich, med. Wochenschr., 1889, p. 426.  
 Hertel, Trans. 28th Heid. Oph. Soc., p. 153.  
 Hirschberg, C. f. p. A. VIII, p. 2. C. f. p. A. XIV, p. 322.  
 Jores, Wesen und Entwicklung d. Arterioskler., Wiesbaden, Bergmann, 1903.  
 Klotz, Adami in Am. J. of Med. Sci., Oct., 1909.; Jour. Exp. Med., 1906, p. 323.  
 Knapp Discussion T. O. S. XXIV, 1904 p. 172.  
 Leber, Graefe-Saemisch, V, 1, p. 663.  
 Magnus, A. f. O. LX, 1874, p. 38.  
 Mannhardt, K. Wochenschr. f. A. XIII, p. 132.  
 Marchand, Eulenberg's Real Encyclopedia.  
 Marple, Med. Record, Mar. 16, 1907; Med. Rec., Mar. 11, 1893.  
 Michaelsen, C. f. p. A., 1899, p. 106.  
 Michel, Lehrb. der Augk., II, Aufl., p. 447. Zeitsch. f. A., VII, p. 5. Z. f. A., XIII, p. 308.  
 Mott, Albutt and Rolleston's System, Vol. VI.  
 Raehlmann, Zeits. f. kl. Med. XVI, 5 u. 6, p. 1. Fortschr. d. Med., 1899, pp. 229-303. Kl. M. f. Augk., XXVII, p. 242.  
 Schleich, Mitteil. aus d'Oph. Klinik in Tübingen II, 2, p. 202.  
 Scultet, Ref. in Wilbrand und Saenger, Vol. IV, p. 175.  
 Seydel, A. f. A., XXXVIII, p. 157.  
 Sidler-Huguenin A. f. A. LI, p. 27.  
 Thoma, A. f. O. XXXV, 2, p. 1.  
 Tyson, Brights and Diabetes.  
 Virchow, Über d. Clinic u. aname in Gefasseaparat, Berlin, 1872.  
 Von Graefe, Angiektasie, Leipzig, 1808, p. 33.  
 Wilbrand und Saenger, Path. d. Netzhaut, IV, Wiesbaden, 1909, p. 248.

### STEREOGRAM 32. RETINAL HEMORRHAGE IN ARTERIOSCLEROSIS.

Right eyeground of a man 47 years of age. Works in a brewery where he performs heavy manual labor. Consumes a great quantity of beer daily. Blood pressure, 190 mm. Hg. Urine contains albumin and casts, but the specific gravity, quantity and urea content do not indicate advanced degeneration of the kidneys. Central vision in the right eye was suddenly lost about one month ago. In the left eye vision is normal. Eye tension is not unduly increased. Ophthalmoscopic examination reveals extensive degeneration of the retinal vessels in both eyes. *Right eye:*—Physiologic







Fig. 69.

Linear Hemorrhage in Nerve Fiber Layer of Retina (shown at a).



Fig. 70.

Hemorrhages involving the Deeper Layers of the Retina.





Fig. 71.

Figs. 71 and 72—Hemorrhages Involving the Layer of Rods and Cones, penetrating the External Limiting Membrane and Tending to Spread between the Retina and Choroid.



Fig. 72.



Fig. 73.

Fig. 73—Small Hemorrhagic Extravasation Elevating but not Rupturing the Internal Limiting Membrane of the Retina, near the Macula.

excavation of the optic disc, but no neuritis. The macular and supratemporal regions contain numerous small, striate hemorrhages. Most of the extravasations are recent, but a few exhibit yellowish-white patches of partial absorption and degeneration. The bleeding has occurred principally along the course of the superior temporal vessels, but no special point of closure appears in the vein. No exudates are seen in the fundus. The retinal arteries are irregularly narrowed and bordered by distinct, white lines. Several small, white plaques of degeneration appear on the surface of the inferior temporal artery. A crossing artery indents and obscures an underlying vein in the manner characteristic of arteriosclerosis. The veins are white bordered, tortuous and alternately constricted and dilated. The venous terminals are unduly prominent and very tortuous. The inferior temporal veins are wider at the periphery than at the disc. This case might well be classed as one of hemorrhagic albuminuric retinitis.

Inasmuch as bleeding is a prominent feature in most of the retinal diseases later to be described, the text will be better understood if, at this point, the subject of hemorrhage is somewhat fully discussed.

RETINAL HEMORRHAGE is one of the most common pathologic conditions observed with the ophthalmoscope and occurs as a symptom in a variety of local and general diseases. The amount of blood extravasated varies from minute extravasation up to an almost continuous sheet covering the fundus. Extensive bleeding breaks down all barriers, but when moderate its position in the retina usually can be determined by its form. Thus, in the nerve fiber layer the blood follows the direction of the fibers and the hemorrhages appear linear, striped, tongue-shaped or as though splashed on with a brush (Fig. 69). In the deeper layers the extravasations are rounded or irregular, their form being determined by the perpendicular fibers of Müller which prevent the blood from spreading (Fig. 70). If blood penetrates the external limiting membrane of the retina and appears among the rods and cones, it tends to spread between the choroid and retina (Figs. 71-72). Multiple capillary hemorrhages consist of small, round, disseminated flecks, distributed independently of the position or direction of the retinal vessels (Stergm. 38). Small, round or oval extravasations, which lift but do not rupture the internal limiting membrane, occur especially in the macular region (Fig. 73). Allied to this form are the preretinal hemorrhages. If hemorrhages are very large or the blood is extravasated with rapidity and force, it enters the vitreous. Bleeding into the perivascular sheaths may appear as stripes along the walls, or as a smooth outlined swelling on or around

the vessel. These hematomata are distinguishable from vascular ectasiæ by their dark color and absence of axial light streak. When first extravasated, arterial blood may be lighter in color than venous blood, but all hemorrhages soon darken. The color of the blood may be lighter in severe anemias, but this is of little value in diagnosing the nature of retinal hemorrhages. The source of hemorrhage is more readily determined by its proximity to a vessel than by its color; but even this test is unreliable, inasmuch as superficial extravasations may change position or blood may be drawn from a distance into the sheath of a large vessel, or a large capillary hemorrhage may occur anywhere.

The size of a vessel does not determine the amount of blood extravasated. Bleeding from a large vessel may be trivial, while in retinal apoplexy the blood comes from the venules and capillaries.

Retinal hemorrhage is immediately followed by resorption of the blood. Small surface hemorrhages fade away in a week or two, clearing from the margin. Large hemorrhages clear from both periphery and center. White patches composed of phagocytic leucocytes, fatty products of degeneration and dead retinal elements appear in and around the extravasation. The blood, fat and debris are removed through the perivascular lymph spaces and angle of filtration in the anterior chamber. If the retinal vessels are sclerosed, absorption will be slow. The result is prolonged action of the decomposing blood toxins upon the iris and pectinate ligament and this may excite iritis or glaucoma. When hemorrhages are small and confined to the nerve fiber layer, they disappear, leaving no trace. When they are large and deeply situated, yellowish or white discolorations usually remain. When blood has penetrated into the deep layers of the retina, tissue destruction is excessive and the blood clot is replaced by a shining, white cicatrix or by a mass of fibrous tissue. As a rule, little or no pigmentation follows retinal bleeding. When present, it may be either hematogenous or produced by massing and proliferation of the pigmented epithelium. In some cases pigment appears on the disc after absorption of blood.

The functional disturbance produced by a retinal hemorrhage depends upon its situation. If it occurs at the macula, useful vision may be abolished (Fig. 72). If peripherally situated, it may create so little disturbance as to escape observation. Moderate bleeding into the nerve fiber layer does little permanent injury, but in the rod and cone layer it is destructive. Hemorrhage into the vitreous produces cloudy vision, the density of which depends upon the amount of blood in front of the retina.

Under any circumstances, spontaneous retinal hemorrhage is a danger signal of the first importance. Frequently, the first sign of serious disease is a single, small retinal extravasation.

**ETIOLOGY OF RETINAL HEMORRHAGE IN GENERAL.** The etiology of spontaneous retinal bleeding includes all diseases or conditions which greatly increase or diminish the blood pressure, retard or accelerate the circulation, induce changes in the heart or blood vessels or modify the composition of the blood. Foremost are degenerative changes in the blood vessels which occur alone or as the prime pathological factor in other diseases.

**ARTERIOSCLEROSIS.** According to Amman bleeding at the macula is common in senile arteriosclerosis. Occasionally, the ophthalmoscopic picture of an angiopathic retinitis consisting of exudation and hemorrhage may develop in uncomplicated angiosclerosis. Straub followed for six years the courses of 15 cases of retinal hemorrhage from arteriosclerosis. The bleeding exhibited a strong disposition to occur in the left eye. Straub states that the prognosis for life is not so bad in the young and very old as it is in people of middle life. For example, nine of the cases were between the ages of 45 and 65 years. Of these, six died within six years, five of them from apoplexy. Derby observed 90 cases of retinal hemorrhage in people over 40 years of age. Of these he ascertained the death of 31, of which 25 died within two years; 14 from apoplexy and 11 from heart disease. Hemorrhage from the retinal vessels is not uncommon in degenerated eyes. I have examined microscopically a number of such bulbs in which excessive hemorrhage had occurred, either spontaneously or from slight injury.

Retinal bleeding usually depends upon degeneration of the blood vessels. The blood may escape either by rhexis or diapedesis. Harms does not accept the view that the sclerosed parts of a vessel tend to bleed, although he admits that they easily rupture from sudden fall of intraocular pressure, as in opening the anterior chamber. A sclerosed wall offers greater resistance to gradually developed pressure than a normal wall, and diapedesis occurs less readily. The unyielding character of sclerosed vessels is demonstrated by the following experiments of Thoma on cadavers. In an arteriosclerotic body, four liters of salt solution, injected into the blood vessels, produced edema of the lower extremities, while in non-sclerotic subjects seventeen liters were injected before edema appeared. A much longer time was required to inject a given amount of fluid into an arteriosclerotic subject than into one with normal vessels.

In *leukemia* and *pernicious anemia* hemorrhages into the retina occur in nearly all cases. In leukemia fresh extravasations often contain white centers. Similar hemorrhages occur in pernicious anemia but it is not certain that the white centers appear at the time the blood is extravasated. The peculiarities of leukemic hemorrhages are considered in connection with the retinitis that occurs in that disease (Stergm. 39).

ANEMIA. Retinal hemorrhage is a doubtful event in, so-called, *simple anemia*. Anemias always are secondary, therefore, associated retinal hemorrhage should be classed as a symptom of the underlying disease. Pick found retinal hemorrhages in 30 per cent of the *intense* anemias produced by carcinomatosis.\* The anemic condition may be responsible for retinal bleeding which occurs in cases of intestinal parasites,† tuberculosis, exhausting hemorrhage, etc.

CHLOROSIS. In chlorosis as in simple anemia, retinal bleeding is infrequent. When it does occur other complications usually coexist, *i. e.*, disordered menses, or choked disc. Kaufmann frequently has found fatty degeneration of the intima in the blood vessels of chlorotics. Thrombosis, as a rare event, belongs to the history of chlorosis. Neither retinal hemorrhage nor optic neuritis should be attributed either to anemia or chlorosis until all other possible causes have been eliminated. It is a point of diagnostic importance that in pernicious anemia hemorrhage into the retina occurs in all cases, while in simple anemia and chlorosis it is extremely rare.

MALARIA. Malaria appears to be a frequent cause of retinal bleeding in those regions where the disease is severe in type. The hemorrhage may be slight or so extensive as to appear in the vitreous. One or both eyes may be affected. It may occur in the first week during the fever or in the late, cachectic stage, at which time vascular disease may exist. In ten cases that died from pernicious and comatose forms of malaria, Guarnieri regularly found stasis, edema and hemorrhage in both brain and retina. In the capillaries the red blood corpuscles contained plasmodia and pigment and the white cells enclosed pigment.

SYPHILIS. Simple retinal hemorrhage is uncommon in syphilis. When present it generally occurs in connection with papillitis, retinitis or local vascular disease.

STASIS. When the retinal vessels are sound they withstand a high degree of distension without loss of blood. This is well illustrated in cases

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\*Also Stephen Mackenzie.

†*Ankylostomum duodenale* (Nieden, Fisher and others.) *Bothriocephalus latus* (Natanson and others).

of retinal cyanosis (Stergm. 58), in which bleeding occurs only in late stages after vascular degeneration has ensued. Cases are reported of retinal hemorrhage due to venous congestion produced by paroxysms of whooping cough\* and epilepsy.† In these and similar cases, vessel alterations cannot be excluded simply because they are unrecognizable with the ophthalmoscope. It is doubtful if any degree of muscular exertion produces sufficient congestion to cause retinal hemorrhage. I have microscopically examined the eyes of six healthy adults killed by the application of a powerful electric current, but in none of them did I discover retinal bleeding, although by this method of death all the voluntary muscles are thrown into violent contraction. A condition known as *traumatic asphyxia* ensues when the body, especially the thorax, is subjected to great external pressure. In this condition bleeding into the retina and optic nerve may occur, sometimes several days after the compression.

NATAL RETINAL HEMORRHAGE is produced in a somewhat similar manner from compression of the head and thorax during birth. Possibly, attempts to breathe while the body is yet in the parturient canal may induce sufficient turgescence to cause retinal bleeding. These hemorrhages are said to occur in nearly all cases of protracted and complicated labor and in feeble children. Thus, Coburn in 37 stillborn and short-lived infants, found retinal hemorrhages in 17. Natal hemorrhages usually disappear leaving no trace of their existence in the retina. Naumhoff suggests that, occurring in the macula, they are the usual cause of congenital amblyopia. Local stasis from obstructed venous outflow is the principal cause of hemorrhage in choked disc, thrombosis of the central vein and compression of the nerve. Sudden local congestion may be the determining cause of bleeding from diseased retinal vessels, as when the aqueous is evacuated in operations for glaucoma. Besides the diseases already mentioned, hemorrhages in the retina have been reported as occurring in influenza, typhus, typhoid, plague, erysipelas, smallpox, measles, scarlet fever, gout, septic retinitis, Weil's disease, childbirth, climacteric, intestinal disorders, menstrual disturbance, masturbation, sexual excesses, etc. Retinal hemorrhage is a rare and inconspicuous event in ocular disturbances produced by poisons. When bleeding occurs it appears to be associated with retinal and nerve congestion, indicating that it is due to changes in the circulation and walls of the vessels rather than alterations in the composition of the blood. Poisoning by coal gas, snake venom, nitrobenzol, analin, phos-

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\* Landesburg, Teillais.

† de Gouvea.



phorous, sulphuric acid, felix mas and alcohol are described as having produced extravasations of blood in the retina. Wagenmann described a case of extensive skin burn in which multiple, small retinal hemorrhages appeared in both eyes. They were attributed to the same blood changes which after severe burns produce ulcerations in the bowels.

There are two forms of intraocular bleeding which present features requiring extended descriptions. These are, *preretinal hemorrhage* and *intravitreous hemorrhage*.

PRERETINAL or SUBHYALOID HEMORRHAGE appears in the form of a large, thin, discoid clot on the surface of the retina. After a few days the blood corpuscles separate and form a lower, dark-red part, divided by a sharp, horizontal line from an upper, pale-red part composed of plasma. The plasma is resorbed before the corpuscles, leaving a slight opacity. Instead of being resorbed the blood may enter the vitreous. Griffiths has seen the vitreous suddenly fill with a broken-down blood clot from yielding of the structures in front of a subhyaloid hemorrhage. It is generally assumed that these hemorrhages are situated between the retina and vitreous. Were this true it might be expected that the blood would be diffusely distributed and gravitate to the bottom of the globe; whereas, the clots are sharply circumscribed, present great uniformity in shape and manifest little tendency to change their position. These characteristics render it probable that the blood is immediately beneath the internal limiting membrane of the retina. Probably, the clouded appearance which remains after resorption of the plasma is an opacity in the retina. A preretinal hemorrhage does not injure the perceptive tissue of the retina; consequently, after removal of the blood, vision will be completely restored.

STEREOGRAM 33. PRERETINAL (SUBHYALOID) HEMORRHAGE. Woman 29 years of age. She is an ill-nourished, sweat-shop seamstress. About one month ago, while bending over her work, a cloud suddenly appeared before the left eye which completely destroyed central vision. With the ophthalmoscope, an enormous extravasation of blood is seen covering the entire macular region. It lies on the surface of the retina in front of the vessels, is slightly elevated and its outline is sharply defined. The lower portion of the hemorrhage consists of a dark-red clot and the upper portion of very light-red plasma. The retinal vessels present no positive evidence of arteriosclerosis. Physical examination fails to demonstrate tuberculosis or syphilis.

At the time this case came under observation tuberculin was not employed for diagnostic purposes.

**INTRAVITREOUS HEMORRHAGE.** Blood in the vitreous appears ophthalmoscopically as slightly movable flocculi or ill-defined masses; sometimes as a large clot the surface of which may undulate on movement of the eye. These hemorrhages appear black in color or give a reddish gleam. When situated far forward in the vitreous and the pupil is widely dilated, the effusion may be recognized as blood by oblique illumination. By transillumination, fresh blood is transparent but clotted blood casts a shadow. Sometimes the iris is colored brown or green from absorption of blood pigments. Months usually elapse before resorption of a vitreous clot is completed. The blood may color the vitreous and cause red vision, which later becomes yellow or green. Fuchs has observed in some cases of traumatic vitreous hemorrhage, some time after the injury, the coloring matter of the blood clot suddenly dissolve and stain the ocular fluids. In such cases the aqueous is colored red and the iris appears as though seen through ruby glass. When hemorrhage has been large and the vitreous badly disrupted, permanent opacities may remain or membranes of dense connective tissue may form which seriously impair vision (Stergm. 61).

In profuse retinal hemorrhage from any cause, blood may enter the vitreous. Aside from such incidental cases, a special form occurs known as *recurrent intravitreous hemorrhage in young people*. Von Graefe first separated this class of cases in which vision is periodically lost from recurring hemorrhages into the vitreous. Subsequent observations of this affection have added to our knowledge of its clinical features but have done little to clear up its etiology and pathology. It occurs in young people, chiefly males. Suddenly, without apparent cause, vision of the affected eye is lost from effusion of blood into the vitreous. As a rule, the amount of blood extravasated is sufficient to obscure the eyeground. Exceptionally, blood appears in the anterior chamber. Resorption quickly ensues, but after a variable period hemorrhage recurs and vision again is lost. The attacks may be few in number or repeated until eyesight is destroyed. As in other forms of intraocular hemorrhage, iritis or glaucoma may result. Many cases give a history of recurring epistaxis which ceases after the advent of intravitreous bleeding. The hemorrhages usually appear first between the fifteenth and twenty-fifth years, very rarely after the fortieth year. One or both eyes may be affected. When unilateral, a marked predilection is shown for the left side. Both eyes may be affected simultaneously or at different times. The mere presence

of blood in the vitreous does not increase intraocular tension,\* and in some cases tension is diminished.† The interval between hemorrhages varies from a few weeks to months. In rare cases, which may belong to a different class, years elapse between attacks.‡ Considering that the vitreous contains no blood vessels, resorption in this form of bleeding is rapid, two to four months being the usual period. A remarkable case is reported by Jacquesu in which the hemorrhage was removed in three days. In this case it is probable that the blood came from the ciliary body and lay between the lens and vitreous. Exceptionally, resorption is delayed for a year or longer.§ The predominance of male over female cases is shown by Wilbrand and Saenger, who collected 73 typical cases of this affection of which 15 were females and 58 males.|| Among females it is possible that the periodic loss of blood from menstruation may diminish the tendency to intraocular hemorrhage.

The source of the hemorrhage is a matter of doubt. Probably it is not the same in all cases. The occasional association of changes in the choroid has led many authors to regard this membrane as responsible for the bleeding.¶ In the few microscopic examinations which have been made in cases of recurrent intravitreal bleeding, vascular changes have been reported.\*\* A combined disease of the choroid and retina is assumed by Leber to be responsible for these hemorrhages. Michel makes the important statement that a vitreous hemorrhage often is the first symptom of tuberculosis of the ciliary body. I have seen one case in which the bleeding was due to a small tubercular lesion, situated at the extreme anterior limit of the ophthalmoscopic field. The uniformity of symptoms exhibited by cases of this affection indicates a common cause, regardless of the source of bleeding. Axenfeld and Stock affirm that hemorrhage due to tuberculosis of the vessel walls may occur without evident lesion of the iris or choroid. If this is true, it explains how blood may come from the ciliary body, choroid or retina without giving visible evidence of its origin. The characteristic, sudden, abundant hemorrhage that occurs, suggests rupture of a single, large vessel, differing from the slow bleeding of hemorrhagic retinitis, in which a lake of blood may cover the fundus without escaping into the vitreous (Stergm. 46).

\*Zieminski, five cases in which tension was unaffected.

†Nieden, tension diminished.

‡Bening, interval 11 years. Kipp, interval 6 years.

§Mayweg, 18 months.

||Bening, 217 cases; females 74; males 143.

¶Nieden, Fehr, Knapp.

\*\*Scheffel, four cases. Fehr, three cases.

From the evidence at hand, it would appear that recurrent intravitreal hemorrhage is due to some process, probably tubercular, which erodes a large vessel in the ciliary body, retina or choroid.

Other diseases which have occurred in connection with intravitreal hemorrhage and, presumably were causative, are hereditary and acquired syphilis, gout, menstrual diseases, anemia, exhaustion, malaria, endogenous infection, constipation, hypertrophy of the heart, hemophilia, purpura and scurvy. When the tendency to hemorrhage already exists, its immediate cause may be congestion due to stooping, sneezing, etc.

The prognosis of intravitreal bleeding appears to be more favorable in the young than in those of advanced years. If iritis develops it may subside with disappearance of the blood, but it is a serious complication and, if in addition eye tension is increased, the eye will probably be destroyed by glaucoma.

### *Bibliography.*

- Amman, Beitr. z. p. Augk. XXXVIII., p. 1.  
 Bening, Inaug. Dis. Giessen. 1900.  
 Coburn, A. of Oph. 1904, XXXIII. p. 256.  
 De Gouvea, Annal. d'Ocul. CXVIII, p. 96.  
 Derby, Mass. Med. Soc., June 8, 1907.  
 Fehr, C. f. p. A. XXV, 2.  
 Fisher, C. f. p. A. 1896, p. 366.  
 Fuchs, 3d Ammer. 1908, p. 470.  
 Griffiths, Norris & Oliver, Vol. 3, p. 383.  
 Guarnieri, Arch. p. l. s. med. XXI, p. 1.  
 Harms, A. f. O. XXXIV, 1905 & LXI, 1905. Corespbl. f. Schw. Artz, XXVIII, 1908, p. 338.  
 Jacques, Oph. Klin., 1899, XXII.  
 Jarnatowski, Recueil. d'O., p. 577.  
 Kaufmann, Lehrb. d. sp. path. Anat., p. 45.  
 Kipp, Trans. A. Oph. S. 1895, p. 435.  
 Landesburg, Med. & Surg. Rep. XLIII, Sept.  
 Lawford, Oph. Rev. 1893, p. 347.  
 Mayweg, Berich u. d. Oph. Gel. z. Heidg., p. 92.  
 Natanson, Jahreber. f. Oph. 1894, p. 528.  
 Naumhoff, A. f. O. XX XVI, 1890, p. 180.  
 Niden, 14 Sitz. d. Heidg. Oph. Gesel. p. 8; Cent. f. p. A. 1903, p. 366.  
 Pick, Klin. Monsb. f. A., XXXIX, p. 177.  
 Scheffels, Deutsch. Med. Wochensft. 1897, XIII.  
 Straub, Klin. M. f. A. XLVI, 1908, p. 517.  
 Teillias, Rec. d'O. 1895.  
 Thoma, quoted by Mott, Allbutt & Rolleston, vol. VI.  
 Von Graefe, A. f. O., vol. 1.  
 Wagenmann, A. f. A. XXXIV, 2, p. 111.  
 Welch, Trans. Ass. Am. Phys. 1897, p. 121. Allbutt & Rolleston, 1909, vol. VI, p. 762.  
 Wilbrand & Saenger, Path. d. Netz. 1909, p. 215.  
 Ziernski, Rec. d'O. 1887, p. 709.

## CHAPTER VI.

### ANGIOPATHIC RETINITIS.

Simple sclerosis of the retinal vessels is never a local disease, and the slightest pathologic change in the vessel walls is positive evidence that wide-spread vascular disease is imminent or already established.

Ophthalmoscopic evidence of angiosclerosis varies from mere tortuosity of vessels to destruction of the retina by hemorrhage, exudation or degeneration. The intimate relation between degeneration of the blood vessels and chronic inflammation of the inner layers of the retina has led to the belief that symptomatic retinitis, as it occurs in different diseases, is only an expression of disturbed circulation.\* The familiar ophthalmoscopic picture of "Bright's" disease, formerly supposed to characterize that affection, has been known to occur without albuminuria as the result of infection in scarlatina, measles, erysipelas, malaria, anchylostoma and syphilis. Typical examples have been reported also in diabetes, scurvy, pernicious anemia, carcinomatosis, brain tumor, in poisoning by lead and felix mas; and in the absence of known toxins or infection, in uncomplicated arteriosclerosis. Wilbrand and Saenger, and Parsons have each reported cases exhibiting a "macular star" for which no cause could be found. While the fundus changes in any case of symptomatic retinitis may strongly suggest the nature of the general disease, they cannot be considered as strictly diagnostic. Therefore, it is inaccurate to describe special forms of retinitis as characteristic of a particular disease. To meet a difficulty in terminology, Wilbrand and Saenger designate all inflammations of the inner retinal layers "*angiopathic retinitis*" and qualify this term by the name of the associated disease, as "albuminuric angiopathic retinitis," "diabetic angiopathic retinitis," etc. This nomenclature may be not wholly free from criticism, but it possesses the great advantage of convenience. Although the appearance of an angiopathic retinitis is not always diagnostic, it usually suggests the nature of the

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\*A theory has been advanced that both vascular and tissue degeneration are caused by the local action of some toxin. In the retina, tissue degeneration appears to be secondary to mechanical closure of vessels.

general disease. Therefore, from a clinical standpoint it is desirable to separate and describe angiopathic retinitis as it usually appears when associated with certain diseases.

**ANGIOPATHIC ALBUMINURIC RETINITIS.** A distinction is drawn between the acute and chronic forms of retinitis occurring in albuminuria. The *acute* type occurs in acute diffuse nephritis, in which a stage of retinal hyperemia and exudation often is followed by recovery. In the *chronic* type the stage of hyperemia almost invariably passes into that of degeneration. These two forms are known also as the *exudative* and the *degenerative*.

In the great majority of cases angiopathic retinitis occurs in connection with chronic nephritis. The ophthalmoscopic picture may present any pathologic condition of the retina resulting from vascular degeneration. Clinically, however, the term "albuminuric retinitis" has been applied to a group of symptoms consisting of venous distension, mild neuritis, retinal opacity, exudation, hemorrhage, retinal degeneration and atrophy. Any or all of these changes may exist in a given case. The features presented by the average case of angiopathic retinitis are best described by dividing the disease into the following four stages: (1) Precursory or Prealbuminuric; (2) Hyperemic or Congestive; (3) Degenerative; (4) Atrophic.

**STEREOGRAM 34. PRECURSORY STAGE OF ANGIOPATHIC RETINITIS.** Left fundus oculi of a laborer 53 years of age. Addicted to the immoderate use of alcohol and tobacco. Blood pressure 190 mm. Hg. The urine contains neither albumin nor casts, but the percentage of urea is low. Has persistent, dull headache. The fundus conditions are similar in each eye. The optic disc has a brick-red color. The retinal arteries pursue a straight course. Their axial light streak is very bright and well defined, producing the effect known as "silver wire arteries." The walls of the arteries are not visible, but that they are sclerosed is demonstrated by the manner in which they indent underlying veins. The larger veins are widened and tortuous and their light streaks are pronounced. The small vessels around the disc and the small veins throughout the fundus are unduly distinct and their terminals twisted.

The class of fundus cases here defined as precursory, or prealbuminuric retinitis are only examples of retinal arteriosclerosis which have just entered the stage of arterial rigidity and venous dilation. The systemic angiosclerosis which invariably attends these retinal changes usually results in chronic nephritis. In a certain number of cases, however, vascular degeneration and its sequelæ are farthest advanced in the brain,

heart or other organs, and the focus of disease is situated elsewhere than in the kidneys. Of all phases presented by angiopathic retinitis, this is the most important to recognize. The spectacular fundus pictures of the later stages of this disease indicate that irreparable destruction has already occurred and a fatal termination is impending; whereas, if the slight but significant alterations of the precursory stage are discovered, the progress of disease may be arrested or, at least, delayed. The important retinal vascular changes of the precursory stage frequently escape observation. They are not prominent and must be diligently sought with the ophthalmoscope through a wide pupil.

The effect of heightened blood pressure on the retinal vessels is manifested by the veins which become visibly distended and tortuous. The condition of the terminal venous twigs is of especial diagnostic significance. They present a curly, "corkscrew" appearance, described also as "crinkled" by Alleman. These little vessels should always be looked for around the macula and disc. De Schweinitz calls attention particularly to one of them, frequently seen descending from a transverse macular artery. The venous distension which exists in established arteriosclerosis extends to the capillaries on the disc, and the latter assumes a dirty, brick-red color. Papillitis may develop in the absence of albuminuria, from uncomplicated vascular degeneration (Stergm. 31).

**DIAGNOSIS.** A fundus exhibiting the features of established angiopathy is "prealbuminuric" in that this condition frequently antedates the development of nephritis and always precedes albuminuric retinitis. The case presented above is typical, with straight arteries and tortuous veins. In another case it may happen that the arteries have retained more or less of the tortuosity acquired in the stage of arterial relaxation and dilation. The characteristic indentation and obscuration of a vein by an overcrossing artery are positive ophthalmoscopic proof of retinal arteriosclerosis. Next in diagnostic importance are crinkled venous terminals and a strong axial light streak on the arteries. In a doubtful case, a retinal hemorrhage, however trifling, or an opaque speck on an artery would be conclusive evidence of angiopathic disease. In hypermetropia the disc may be red and the vessels tortuous, but there is no vascular degeneration.

**DEVELOPED RETINITIS. SYMPTOMS.** In nearly all cases of angiopathic albuminuric retinitis both eyes are affected.\* Frequently, the two

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\*Vascular degeneration probably exists in both eyes in all cases of retinitis of albuminuria.

eyes are attacked at different times and in varying degree. Moreover, the manifestations of the vascular disease may differ widely in each eye. Thus, in case of stereogram 43 typical angiopathic retinitis existed in the right eye and closure of the central artery in the left. All the external ocular manifestations of arteriosclerosis may appear in albuminuria, especially ecchymoses and fugitive edema of the eyelids. Wharton Jones and Schöbl have seen hemorrhage into Tenon's capsule. Post-hemorrhagic glaucoma may develop, but spontaneous glaucoma is unusual, notwithstanding the fact that, in chronic albuminuria, degeneration of the blood vessels is extreme. No connection has been traced between albuminuria and development of cataract. Detachment of the retina is common in the late stages of chronic nephritis, especially if there is anasarca. In chronic nephritis the vessels of the choroid undergo degenerations analogous to those occurring in the retina, but the disastrous results of retinal arteriosclerosis usually are averted in the choroid by the free anastomoses existing between its vessels. It is said, however, that chorioretinitis of angiopathic origin can develop. The presence of this complication would be recognized by pigment heaped around white plaques of degeneration and by disturbance of the retinal epithelium and pigment wandering.\*

The degree of functional eye disturbance experienced in the retinitis of albuminuria depends upon the nature and location of the lesion rather than upon the severity of the retinal inflammation or gravity of the general disease. Probably, visual acuity always is blunted even though it may not be indicated by the ordinary card tests. At first, there is complaint of a slight fog before the eyes, an effect which is produced by the presence of retinal opacity before the percipient layers of the retina. Lesions situated in the macula or detachment of the retina produce great impairment of vision. Hemorrhages affect sight according to their situation and extent. Total loss of vision seldom, if ever, is due to uncomplicated retinitis or subsequent atrophy. Sudden blindness in both eyes means uremic amaurosis. Occurring in one eye it may indicate either retinal detachment or closure of the central artery. The visual fields usually correspond to the destruction wrought in the retina, differing in this respect from diabetes in which amblyopias with scotomata occasionally occur. Concentric limitation of the fields, described by Schlesinger, and paracentral ring scotoma by Leber, have not been accepted as belonging to the history of the disease. Color scotomata have been described by Simon and others as characteristic

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\*The condition is so rare that complications probably exist.



of albuminuria, a conclusion which has not been confirmed. These scotomata present wide variations and probably depend upon macular changes. Cerebral hemorrhage is not uncommon in albuminuria, and hemianopsia or other central disturbance of vision may ensue. Ophthalmoscopically visible changes exist long before vision is affected or albumin appears in the urine, hence retinitis sufficient to attract the attention of the patient is indicative of advanced degeneration of the kidneys. It is only in the early period that information acquired through use of the ophthalmoscope may be the means of preserving life. In many cases of contracted kidney albumin is not demonstrable in the urine, a fact which explains many instances of retinitis reported as occurring in healthy subjects.

**HISTORY AND COURSE.** It is difficult to estimate the frequency of retinitis in chronic nephritis. From a collection of 935 cases of the acute and chronic types, Parsons found the average ratio of retinitis to be 22.4 per cent. This percentage represents mostly cases of well developed retinitis, but fails to include a much larger number in which minor vascular changes existed. Nettleship collected 43 fatal cases in young adults, of which 33 had retinitis.

As regards sex, nephritic retinitis appears to be about twice as frequent among men as among women. Parsons found in 180 reported cases, 68 per cent males and 32 per cent females.

Retinitis in Bright's disease occurs at all periods of life; the usual age is between 30 and 60 years. The youngest case reported is by C. S. Bull, in a child 5 years of age. Another, aged 9 years is reported by de Schweinitz.

Retinitis may occur with any form of nephritis, but is most frequent in contracted kidney. It is uncommon with the large white kidney and more rare in the acute nephritis of pregnancy and scarlatina.

The course of the disease is eminently chronic. Degeneration of the retina and deterioration of vision keep pace with the advancing nephritis. Arrest or subsidence of the process in chronic cases must be extremely rare, although instances have been reported by Admuck, Steffan and others. Temporary improvement in vision may be due to local causes, but cure cannot be expected while the nephritis continues. Uremic attacks may be attended by temporary or permanent amaurosis.

The prognosis as to life is extremely unfavorable in fully developed cases of albuminuric retinitis. In nearly all cases death ensues within a few months or years. In 103 cases, C. S. Bull found that 87 per cent died within two years. Prognosis is no better among the young than the old.



Fig. 74.

Fig. 74—Angiopathic. Albuminuric Retinitis. R. retina; C, choroid; S, sclera.  
Edema of the entire Retina.



Fig. 75.

Fig. 75—Angiopathic, Albuminuric Retinitis. Cystoid spaces in the internuclear Layer.



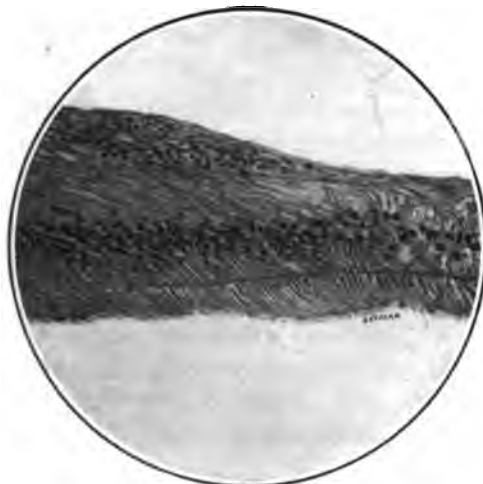


Fig. 76.

Fig. 76—Edematous Infiltration around the Macula, in Albuminuric Retinitis.

Fig. 77—Retinal Discoloration left after removal of Superficial Hemorrhage by Leucocytes.



Fig. 77.



Fig. 78.

Fig. 78—Deep Hemorrhage Replaced by Plaque of Cicatricial Tissue with Pigmented Borders.



Nettleship collected 80 cases of chronic nephritis in subjects under 21 years of age. Of these, 60 came to necropsy. The influence of habit and environment upon the course of the disease is forcibly illustrated by the work of Possauer, whose material was collected from 67,000 cases of all kinds. In cases of albuminuric retinitis among the poor who visited the clinics, all males died within two years, a mortality of 100 per cent. Among women of the same class only 68 per cent died. Among private patients of means, the death rate was 59 per cent for males and 53 per cent for females. Among the poor the longest duration of life after discovery of retinitis was 6 years, and among private patients 11 years. These statistics illustrate the value of good hygienic conditions in the treatment of nephritis.

**PATHOLOGY.** The pathologic anatomy of angiopathic retinitis includes all morbid changes that result from vascular disease. Inasmuch as the retinal blood vessels are a terminal system and the only lymphatics are those which accompany the vessels, any obstruction to circulation is followed by disturbance of nutrition, the visible manifestations of which are edema and hemorrhage. Edema appears first in the nerve fiber and inner reticular layers, but the entire retina soon is infiltrated with abundant fluid (Fig. 74), poor in albumin and readily absorbed. The nerve fiber layers are separated by the transudate, the inner layers being lifted up at the expense of the vitreous space. Cystoid spaces may form in the internuclear layer (Fig. 75). Edematous infiltration is especially marked around the macula (Fig. 76). If edema is long continued the nerve fibers swell and lose their outline. Transient stasis may not injure the ganglion cells but these delicate structures are the least resistant of the retinal elements and quickly succumb in areas insufficiently supplied with blood. Usually, the hemorrhages are superficial, the blood lying between the axons of the nerve fiber layer, where they present the ophthalmoscopic picture characteristic of striate or "flame-shaped" hemorrhages. Blood extravasated into the retina seldom clots. Striate hemorrhages are slowly removed by leucocytes, leaving the retina slightly discolored (Fig. 77). Deep hemorrhages often destroy the entire thickness of the retina which then is replaced by a plaque of cicatricial tissue with pigmented borders (Fig. 78). After resorption of the blood from the retina the hemoglobin may remain and form pigmented deposits, but the pigment around the scars of deep retinal hemorrhages is deposited by the retinal epithelium. In albuminuric and some other forms of chronic retinitis, peculiar cell-like bodies, of inconstant size, are found in the ganglion cell and nerve fiber layers. They are gran-

ular or homogeneous in structure and contain refractile, nuclear-like spots (Fig. 79), the origin and nature of which are unknown. They have been variously regarded as varicose nerve fibers, degenerated ganglion or neuroglia cells and as clusters of degenerated leucocytes. The latter view is at present popular. The signs of active inflammation are wholly lacking in albuminuric retinitis, and the white exudates so conspicuous in the ophthalmoscopic picture are mostly the products of degeneration. They are principally hyaloid in character but fat also is present and, later, cholesterolin. Calcareous deposits are rare. The "star-shape," which exudates at the macula often assume, corresponds to the radial arrangement of Henle's fibers in this region. The vascular changes are those of arteriosclerosis already described. In the atrophic stage (Stergm. 37) the normal structures are largely replaced by new-formed fibrous connective tissue.

Ginsberg has studied the histologic changes in albuminuric retinitis. He found that the retina contained lipoid substances, principally in the sustentaculum. White discolorations of the retina nearly always indicated subretinal effusion and retinal exudation. Vascular changes were comparatively slight, and Ginsberg regards them as unimportant in the production of albuminuric retinitis.

**ACUTE ALBUMINURIC RETINITIS (Exudative Retinitis).** This form of retinitis occurs in the albuminuria of pregnancy, scarlatina and other exanthemata, and from certain poisons, especially lead. It is best known in connection with pregnancy and only in this relation will it be discussed.

The ophthalmoscopic picture of acute albuminuric retinitis is the same as that presented by the chronic form of retinitis during the exudative stage, but minus the arteriosclerotic changes (Stergm. 35). The special inflammatory character of the acute form upon which some authors insist, is not demonstrable with either the ophthalmoscope or microscope. Detachment of the retina is said to be relatively more frequent than in the chronic type. Occasionally, the acute form is followed by a rapidly progressive, chronic retinitis. In such cases chronic nephritis may have preceded pregnancy. The clinical differences between chronic and acute retinitis consist in etiology, development and course rather than in ophthalmoscopic findings. The arteriosclerotic changes of chronic retinitis are not found in the acute disease, but its angiopathic character is suggested by bleeding and, in one case, by closure of the central artery (Groenouw). The fundus changes are not necessarily pronounced. In none of Nettleship's cases was the retinitis severe.

Silex estimates that one case of albuminuric retinitis occurs in every

3,000 pregnancies. In true cases, *i. e.*, those in which chronic nephritis does not antedate pregnancy, preliminary vascular disease appears to be lacking. When retinitis occurs it usually does so in the second half of pregnancy, although it may appear at any time. The view that most cases of retinitis occur in the first pregnancy is shown to be erroneous by Nettleship who found but four primipara among nineteen cases. A woman in her first pregnancy may suffer from nephritis and retinitis, and escape both in subsequent pregnancies.

Prognosis as to life is good, but in most cases vision is permanently impaired. Silcox found but three out of thirty-five cases that regained normal sight. Howe states that, when retinitis develops in the seventh month and abortion is not induced, permanent blindness may be expected. Occurring in the eighth month less than one-half recover. If it does not appear until within the two last weeks of pregnancy, recovery is assured. Abortion may arrest the disease but does not always insure recovery, and progressive failure of vision may continue after delivery. Traces of the disease usually remain in the form either of white spots in the retina, blanching of the nerve or pigmentation around the macula.

Why retinitis occurs in acute nephritis has not been explained. The statement that it is due to a "uremic state" of the blood is not lacking in vagueness. More likely, both acute nephritis and retinitis have a common origin. Acute diffuse nephritis is attributed to the presence of toxic substances in the blood which, in the process of elimination, are brought into intimate contact with the vascular and excretory cells of the kidneys, and thereby injure or destroy these structures. In a similar manner, the same poisons circulating through the retina may induce the endovascular and parenchymatous changes of acute retinitis. The destructive effect of circulating poisons that act upon the vessels of the kidneys and retina need not be ascribed to "selective action," but to the anatomic fact that both structures are supplied by a terminal system of vessels and the evils of obstructed circulation cannot be averted by collaterals. Nephritis and retinitis may be caused by either endogenous or exogenous toxins.

Uremic amaurosis usually occurs in the course of acute nephritis, seldom with chronic nephritis. As a rule, the fundus is normal or nearly so. Sometimes, however, uremic amaurosis develops in connection with retinitis, and then it is of vital importance to recognize the cause of blindness. A typical case of uremic amaurosis commences with rapid loss of vision which in from 8 to 24 hours eventuates in blindness, even light per-



ception, as a rule, being abolished. This state continues for from 10 to 18 hours, when improvement ensues and after about 48 hours vision is restored. In most cases during the blind period the pupils respond to light. During the attack the urine is diminished in amount, and if albuminuria was present it may disappear. When amaurosis develops in pregnancy it frequently is accompanied by eclampsia. In scarlatina it occurs during desquamation. Uremic amaurosis appears to be more infrequent than albuminuric retinitis. Litzman, in 13 cases of uremic eclampsia of pregnancy, found amaurosis in three. The unconscious condition of the patient with eclampsia often hides an existing amaurosis. The prognosis as to vision is good in those who survive the nephritis. Recurring attacks, however, may result in permanent amblyopia. Two principal theories exist as to the origin of the blindness: (1) high blood pressure, which causes edema of the brain; (2) the irritating action of an unknown toxin. The persistence of pupillary light reaction indicates a cerebral lesion above the primary optic centers, presumably cortical.

**EXUDATIVE (Hyperemic: Congestive Hemorrhagic) STAGE.** With the development of chronic nephritis, a retinitis is no longer "precursory." The exudative stage of chronic retinitis which then ensues includes a wide range of morbid conditions, from hyperemia of nerve and retina, to a state of intense congestion with exudation and hemorrhage. With the establishment of distinct, albuminuric retinitis, the disc becomes markedly congested and its outlines are blurred. Usually, it is moderately swollen. The retina becomes edematous and opaque, especially around the disc, where thickest. Exudates appear first as minute, white, fluffy dots in and around the macula. Later, yellowish spots develop in the swollen retina about the nerve. Small, striate hemorrhages may occur among the exudates and even on the disc. Visible vascular changes, if present, may be obscured by the existing inflammation. In other cases the vessels may exhibit advanced degeneration from the first for, although the retinitis is young, the arteriosclerosis is old. In the early stage of retinitis it is not uncommon to find "silver wire arteries," white bordered vessels or other angiosclerotic changes. In any event, the veins are abnormally broad and tortuous and the arteries of normal width or narrowed. As the disease progresses the congestive and exudative processes increase in severity. The engorged and tortuous veins burrow in the swollen retina. Deep hemorrhages occur and in some cases blood is extravasated into the vitreous. Profuse bleeding is regarded as a grave prognostic sign. The

severe symptoms of this stage may develop with such rapidity that the incipient processes escape observation. Sometimes the course of the disease is marked by acute exacerbations.

**STEREOGRAM 35. EXUDATIVE STAGE OF ANGIOPATHIC ALBUMINURIC RETINITIS.** Right eyeground of a man 37 years of age. Has advanced chronic interstitial nephritis. One month ago he first noticed obscuration of vision in both eyes. The fundus of the right eye presents an unusually characteristic picture of angiopathic retinitis in the exudative stage. The nerve is intensely red, its outline is obscured but swelling is not demonstrable with the ophthalmoscope. The arteries possess a very bright central light streak and their walls hide underlying veins. The veins are somewhat dilated and tortuous the tortuosity being particularly marked in the venous terminals. The disc is surrounded by a number of small striate hemorrhages. On the temporal side of the nerve, the retina contains numerous small, superficially situated, fluffy exudates, some of which cover the retinal vessels. The supratemporal vein is surrounded by a large exudation in the center of which is a small extravasation of blood.

**DIAGNOSIS.** That the pathologic process in this fundus is an angiopathic retinitis is indicated by the red nerve, congested blood vessels, hemorrhages and superficial exudates which overlie the vessels. The absence of flat, sharply outlined plaques of degeneration (macular star, etc.), prove that the retinitis has not as yet entered the stage of degeneration.

**DEGENERATIVE STAGE.** This stage is characterized by gradual subsidence of retinal hyperemia and edema. In course of time, the thickened peripapillary area loses its reddish-yellow opacity and the retina appears thin and, sometimes, striped (wrinkled). White spots of degeneration form mostly about the nerve, rarely as far forward as the equator. These spots are irregular in outline, free from pigment and situated on the surface of the retina where they may cover the vessels. In the exudative stage they present a woolly, soft-edged appearance. The degenerative plaques appear flat, hard and sharply outlined. Magnus states that large plaques may be preceded by diffuse opacity of the retina which subsequently becomes white. Additional minute glistening specks may be scattered about the posterior pole. Frequently, a broad white zone forms around the nerve, producing an appearance which has been compared to a "snow-bank." Around the macula there is an increase in the number of white dots which sometimes fuse and form an encircling ring, or they may be arranged in white rows or lines, radiating from the fovea like rays

constituting the "macular star," at one time supposed to be pathognomonic of albuminuric retinitis. The star rays are unequal in length and seldom form a complete circle.

In a certain proportion of cases, optic neuritis is developed sufficiently to justify the use of the term *neuroretinitis*. This combination may appear at any time previous to the stage of atrophy, after which the nerve and retinal tissues become too sclerotic to manifest marked symptoms of inflammatory reaction. In albuminuric cases of retinal venous thrombosis, neuritis is generally pronounced (Stergm. 47). As a rare event in albuminuria, optic neuritis develops without retinitis. In such cases, after subsidence of the swelling, the nerve exhibits the white hue of post neuritic atrophy. The cause of the neuritis is problematical. Probably, it arises from arteriosclerotic changes in vessels distributed to the disc although some cases may be due to cerebral complications.

In rare cases, vascular degeneration in the choroid may produce visible changes in the fundus, opacities in the vitreous or small detachments of the retina, especially around the nerve. Extensive detachment of the retina, not necessarily dependent upon the choroid, also may occur.

The ophthalmoscopic features of the different stages of albuminuric retinitis may appear singly or in the most varied combinations. Cases presenting all the characteristics of any stage are rare. Albuminuric retinitis usually is observed in the degenerative stage in which the white foci are the conspicuous features, especially the macular spots or star.

**HEMORRHAGIC ALBUMINURIC RETINITIS.** This term is applied to a rare condition which nearly always occurs in subjects who have advanced senile arteriosclerosis associated with albuminuria. It is characterized by recurrent disseminated hemorrhages in the retina, and the absence of neuroretinal changes belonging to albuminuric retinitis. The fundus represented by stereogram 32 is a good example of this condition. An analogous type of hemorrhagic retinitis occurs in diabetes also due to vascular degeneration.

In the retinitis of chronic nephritis, vascular degeneration is the underlying pathologic factor. This may cause closure of the central artery or central vein, producing effects which dominate the scene and obscure other evidence of the disease.

**STEREOGRAM 36. ANGIOPATHIC ALBUMINURIC NEURORETINITIS, DEGENERATIVE STAGE.** Right eyeground of a man, 42 years of age, with advanced interstitial nephritis. The retina is passing from the exudative to the degenerative stage of retinitis. The case is complicated by

a rather severe neuritis which serves to increase the tortuosity of the veins and retinal edema. The different steps of albuminuric retinitis which have been described are never sharply differentiated. For example, in this fundus the retinal edema, hemorrhages and fluffy exudates belong to the stage of exudation or congestion, but the stellate figure about the macula and the large whitish patches, suffice to place the case in the degenerative stage. The retinal edema is most conspicuous about the nerve. The outlines of the disc are completely obscured and the nerve head appears as a muddy, red blur. The engorged and tortuous veins burrow into the swollen retina and disappear. The arteries are narrowed and their central light streaks are sharp, bright and irregular. The hemorrhages are striate, indicating that they lie in the nerve-fiber layer. The large whitish plaques are superficially situated, well defined and nonpigmented. The spots cover the arteries when the two are in contact. The "star" does not completely encircle the macula. Its rays consist of short linear and rounded whitish spots arranged in rows. Small, fluffy, edematous spots are seen where two vessels cross. A group of small dots is scattered among the superior temporal vessels.

**DIAGNOSIS.** In the great majority of cases angiopathic retinitis occurs in connection with chronic nephritis. Exceptionally, it develops in other affections attended by vascular degeneration. Occurring in diabetes the ophthalmoscopic picture may or may not present characteristic features. In diabetic retinitis, visible angiosclerosis, optic neuritis and opacity of the retina are comparatively rare. It must be borne in mind that unrecognized nephritis may exist in connection with any disease which exhibits angiopathic retinitis, particularly diabetes. Albuminuric retinitis in the degenerative stage requires differentiation chiefly from chronic choroiditis.

#### ALBUMINURIC RETINITIS.

Occurs chiefly in posterior polar zone.

White patches, superficially situated; overlie retinal vessels; edges never pigmented.

Veins dilated. Arteries normal width or narrowed. Visible angiosclerosis.

#### CHRONIC CHOROIDITIS.

Lesions distributed throughout fundus.

White patches, deeply situated; always beneath retinal vessels; old lesions looked "punched out"; more or less pigment disturbance.

Retinal vessels normal or uniformly narrowed in old cases.

ALBUMINURIC RETINITIS.

Choroidal vessels not exposed.

More or less optic neuritis.

Retinal hemorrhages.

Vitreous opacities are unusual.

CHRONIC CHOROIDITIS.

Some choroidal vessels usually exposed.

Neuritis is very rare.

Hemorrhage is exceptional.

Vitreous opacities common in early stage.

Albuminuric retinitis is differentiated from other angiopathic forms by diagnosticating the associated general disease.

**ATROPHIC STAGE. RETINITIC OPTIC ATROPHY.** In natural sequence, the stage of degeneration is followed by that of atrophy. All signs of retinal opacity, hyperemia and neuritis subside and the red fundus reflex reappears. As in other forms of retinal atrophy, the fundus remains delicately veiled. The white plaques of the degenerative stage become indistinct, changing from white to a dirty grey and finally disappear or remain as patches of scattered grey specks. The last to disappear is the stellate figure around the macula. Degeneration of the retina leads to retinitic optic atrophy, characterized by the yellowish-white color of the disc. If severe neuritis existed in the earlier stages, the disc may now present the white color and peripapillary pigment disturbance of post-neuritic atrophy. Scattered pigment deposits may appear either from choroidal degeneration or from extravasated blood. The blood vessels undergo extreme degeneration and the smaller branches disappear. This ophthalmoscopic picture is very rare for the reason that albuminuric subjects seldom live long enough for the retinitis to reach this stage.

**STEREOGRAM 37. ANGIOPATHIC ALBUMINURIC RETINITIS. ATROPHIC STAGE.** Left eyeground of a woman 29 years of age. Urinary examination indicates advanced cirrhosis of the kidneys. Three months ago, in the seventh month of her third pregnancy, she became blind from retinitis. An abortion was induced and vision was partly restored. Her vision was defective before the last pregnancy. Ophthalmoscopic examination revealed, in both eyes, angiopathic retinitis in the atrophic stage. Considering the advanced stage of retinitis that now exists, it is probable that pregnancy was an intercurrent event in the course of a chronic nephritis. On the other hand, chronic nephritis with retinitis, following an acute attack, sometimes pursues a remarkably rapid course.

The optic nerve presents the yellowish-white color of retinitic atrophy. The fundus, especially on the nasal side, contains several dirty-grey patches

which are interpreted as retinal cicatrices that have replaced the white plaques of the degenerative stage. In accordance with the rule, the last spots of exudate to disappear are those around the macula. In this case, the few which remain may be remnants of a macular star. Just above the macular region are three small, shining, white specks, probably crystals of cholesterine. Several masses of pigment are grouped along the course of a macular vein. These are regarded as of hematogenous origin for the following reasons: (1) they are superficially situated *upon* dirty-yellow patches, such as remain after resorption of blood; (2) the choroid is not exposed; (3) pigment is within, not around the patch; (4) evident relationship of the pigment to a blood vessel. Varied and irregular vascular degeneration is characteristic of primary angiosclerosis as distinguished from the uniform narrowing and obliteration of vessels due to retinal atrophy (Stergm. 44). All the blood columns are either irregularly constricted or widened. The superior temporal arteries exhibit sectional sclerosis. These vessels form a series of fusiform, thin walled dilations alternating with stretches of artery in which the wall is thickened and lumen constricted. The thickened walls of the constricted sections are not visible. It is said that such translucent thickening is due to proliferation of the intima (endarteritis nodosa). The ascending branch of this artery is faintly visible as it passes beneath a vein. This is evidence of thickened arterial walls. The supranasal artery also exhibits sectional thickening and its walls are visible as white lines bordering the narrowed blood column. When vessel walls become visible it is supposed that the sclerosing process has extended to the adventitia. At its bifurcation, each branch of this artery undergoes dilation. Both the ascending and descending retinal arteries hide underlying veins on the disc, and the spaces which here appear to exist between the ends of the veins and arterial blood columns, measure the thickness of the arterial walls. The veins present irregular blood columns. The smaller veins and terminals are tortuous. Both venous and arterial twigs have been extensively obliterated by the general retinal atrophy. On the disc, the sclerosed arteries press on the underlying veins and obstruct the venous flow. This is manifested by local dilation of the veins distal to the points of pressure.

The thin patches of atrophic retinitis could hardly be mistaken for the deep, pigmented lesions of choroiditis (Stergm. 20). Other forms of retinal atrophy, *i. e.*, retinitis pigmentosa (Stergm. 53), *punctata albescens* (Stergm. 54), occlusion atrophy (Stergm. 44), etc., all possess characteristic fundus changes. Moreover, in all these diseases the retinal

vessels undergo uniform narrowing and obliteration while in atrophy following angiopathic retinitis the vessels present the varied degenerative changes of arteriosclerosis.

**ANGIOPATHIC DIABETIC RETINITIS.** The retinitis which occurs in saccharine diabetes presents no ophthalmoscopic feature by which it can with certainty always be differentiated from other forms of angiopathic retinitis. Peculiarities, however, exist which, in typical cases, permit a probable diagnosis. Thus, the visible angiosclerotic changes so prominent in albuminuria are usually, but not invariably, absent in the retinitis of diabetes.\* Optic neuritis and opacity of the retina also are very rare. The frequency of both albumin and sugar in the urine has rendered it difficult to isolate a special diabetic type of retinitis which is free from suspicion of being a case of albuminuric retinitis in a diabetic subject. Retinitis, however, does occur in diabetes in the absence of albuminuria. Hirschberg has separated uncomplicated cases into two classes which may be accepted as typical of saccharine diabetes. These are: (1) Central punctate diabetic retinitis; (2) Hemorrhagic diabetic retinitis.

**CENTRAL PUNCTATE DIABETIC RETINITIS.** In this type there is little or no evidence of vascular disease, optic neuritis or retinal edema. In the region embraced between the superior and inferior temporal vessels are groups of white spots and lines. They are grouped chiefly around the macula, but do not form a star. Smaller collections occur between the macula and nerve. Isolated spots may appear outside the temporal area, or on the nasal side of the disc, or in front of the vessels. They exhibit no tendency to coalesce and large spots are uncommon. The spots and stripes are irregular in outline and their edges may be serrated. Blood is seldom seen on the spots, but everywhere between them are small, superficial hemorrhages. Hemorrhages occur also beyond the area of the spots. Pigment changes are lacking and the vitreous is clear.

**STEREOGRAM 38. CENTRAL PUNCTATE DIABETIC RETINITIS.** Left eyeground of a laborer, 52 years of age. His vision first began to fail seven months ago and now is 20/200 in each eye. The urine contains sugar, 3 per cent; no albumin, no casts and no acetone. Examination of the feces shows marked pancreatic insufficiency. The fundus changes are symmetrical in each eye. The ophthalmoscopic picture is a remarkable example of Hirschberg's central, punctate diabetic retinitis. The optic disc is normal and the retinal vessels are not visibly sclerosed. The

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\*Cases of visible arteriosclerotic changes in diabetic retinitis, reported by Galezowski, Michel, Knapp.

region embraced by the upper and lower temporal vessels is covered by groups of superficially situated, small, white, discrete, sharply defined spots, many of which possess an elongated shape. Scattered between the spots are numerous small striate hemorrhages. A few isolated spots are situated on the nasal side of the nerve.

The patient was kept in the hospital ward for several weeks, during which time no improvement occurred.

**DIAGNOSIS.** This picture, which is extremely rare, probably is characteristic of diabetes, although the several types of angiopathic retinitis are differentiated by physical examination rather than by the ophthalmoscope. The angiopathic character of this retinitis is indicated by the hemorrhages. It differs in appearance from albuminuric retinitis in the absence of neuritis, visible vascular changes and retinal edema.

**HEMORRHAGIC DIABETIC RETINITIS.** This condition appears to be analogous to the hemorrhagic retinitis of albuminuria. It is distinguished by the repeated occurrence of retinal hemorrhage and by the absence of either white spots or visible vascular degeneration. Any form of retinal hemorrhage may occur. When disseminated, the extravasations are said to be rounder and more deeply situated in the retina than in the albuminuric type. Small punctiform hemorrhages in the retina are suggestive of diabetes. Intravitreous bleeding and vitreous opacities are frequent in this type of diabetic retinitis. Posthemorrhagic glaucoma may develop and invariably is destructive.

Under the name of *atypical diabetic retinitis* Hirschberg places a third group of cases in which diabetes is accompanied by typical retinitis pigmentosa, but a causal relationship between the two conditions has not been demonstrated.

Diabetic retinitis is a rare disease, only a few observers having seen a sufficient number of cases to be convinced that it possesses any special characteristic feature. The case of central punctate retinitis described above, so closely corresponds to Hirschberg's cases, that I am inclined to regard the picture as nearly pathognomonic of diabetes. A large proportion of reported cases, however, were ophthalmoscopically indistinguishable from albuminuric retinitis. Even the macular star is not unknown in diabetes, five cases having been reported, three by Hirschberg, one by Hirschmann and one by Hawthorne.

Although the walls of the blood vessels are seldom opaque, diabetic



cases are not exempt from profound vascular disease, as shown not only by hemorrhage, but by cases of thrombosis of the central vein\* and closure of the central artery.†

Lipemia is an occasional complication of diabetes. In such cases the retinal vessels appear filled with a milky fluid. The arteries have a reddish and the veins a violet tint. Uthoff thinks this appearance is produced by the fat globules which float on the surface of the blood column, forming a layer in contact with the walls of the vessels.

Functional disturbance of vision is far greater in diabetes than in albuminuria. Various types of amblyopia occur, with and without retinitis. Amblyopia with sector defects in the field is common. Optic atrophy with concentric contraction has been reported. Schöbl states that diabetics frequently complain of "glimmering" followed by central amblyopia. Affection of the maculo-papillary bundle, indistinguishable from nicotine amblyopia may develop. Mauthner justly regards these cases as due to the use of tobacco and alcohol, as diabetic subjects are very susceptible to the action of these and other poisons. Aside from retinal and nerve affections, vision in diabetics is prone to be disturbed by imbalance of the extrinsic muscles and also by paresis of accommodation and premature presbyopia. Cataract is a frequent event in diabetes.

Retinitis is a late development of diabetes and is to be regarded as an unfavorable symptom, although the prognosis as regards life is far better than in albuminuria. After the development of diabetic retinitis the general and local conditions often remain unaltered for a long period. Nettleship, in 48 cases, found that 60 per cent lived more than two years.

ANGIOPATHIC LEUKEMIC RETINITIS. Leukemia is manifested in the retina either by neuroretinitis or by retinal hemorrhages without other inflammatory phenomena.

LEUKEMIC NEURORETINITIS. In this form the hyperemia usually is of moderate intensity. In some but not all cases, the disease is distinguished by a pale, orange-yellow color of the fundus and by white centered hemorrhages. Sometimes the blood vessels may be nearly white in color the veins being distinguished only by their excessive size and tortuosity. In other cases the vessels, especially the veins, exhibit white borders. Retinal opacity is present, but seldom so pronounced as in albuminuric retinitis. Escape of blood into the retina is one of the most common events in

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\*V. Michel; Hirschberg.

†Knapp; Dedd.

leukemia. The extravasated blood is said to be lighter in color than in other diseases. Fresh extravasations often present a characteristic appearance, consisting of a central white spot surrounded by blood. Generally the hemorrhages are situated in the nerve fiber layer or on the surface of the retina. If bleeding is profuse all layers of the retina or even the vitreous may be invaded. The lesions of leukemic retinitis frequently are limited to the region anterior to the equator and thus escape observation. The neuritis varies greatly in different cases. Although usually moderate in degree, it may be excessive, the nerve projecting like a mushroom into the vitreous. Occasionally the leukemic fundus presents the picture of "albuminuric retinitis."\* Michel has described two cases of retinal venous thrombosis in leukemia.

The pathologic changes of leukemic retinitis are due to the excess of leucocytes in the blood and their accumulation in the tissues. The orange color of the fundus is supposed to be caused by lymphatic infiltration of the choroid. The perivascular sheaths may be filled with lymph cells and appear as white lines bordering the retinal vessels or may even whiten the entire vessel. Similarly, choked disc may result from infiltration of the nerve and nerve sheaths with leucocytes. The bleeding in leukemia is explained by the altered state of the blood. The great excess of the white cells permits them to accumulate along the walls of the vessels, through which they pass with the red cells. Michel attributes the bleeding to venous thrombi. Probably, Saemisch is correct in regarding the white centers of the hemorrhages as composed of white blood cells. Leber's suggestion that they are small lymphomata has not been confirmed. Leukemic extravasations possess the following peculiarity. In the retinal hemorrhages of other diseases, white areas form only in old extravasations and represent the processes of blood resorption and degeneration; while the white center of a leukemic hemorrhage appears when the blood is extravasated. Examples of both types of white spots are found in leukemic retinitis. Microscopic examinations have thrown little light on the structure of leukemic extravasations, inasmuch as only old hemorrhages have been described.

Deterioration of vision in leukemia appears to depend upon actual lesions in the retina and nerve rather than on amblyopias of toxic or cerebral origin. Schmitt-Rimpler, however, has reported a case associated with retrobulbar neuritis.

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\*Schirmer; Kramszky.

Other ocular manifestations of leukemia are lymphoma of the eyelids, exophthalmus due to lymphogenous deposits or to hemorrhage in the orbit, enlargement of the lachrymal gland, iritis, iridocyclitis and hyphema.

**HEMORRHAGIC LEUKEMIC RETINITIS.** In this form of retinitis the signs of congestion are slight or lacking. The fundus may or may not present the yellowish cast of leukemia. Some dilation of the veins usually exists. In the following case the most interesting clinical feature was the presence of a white center in a fresh hemorrhage.

**STEREOGRAM 39. LEUKEMIC HEMORRHAGIC RETINITIS.** Right fundus oculi of a man 32 years of age. Syrian. Entered the clinic complaining only of slight conjunctivitis. He was intensely anemic. Blood examination demonstrated the presence of lymphatic leukemia. Urine normal. Vision 20/20. With the ophthalmoscope, numerous characteristic white centered hemorrhages along the arteries and veins were found in both eyes. Three days after coming under observation, central vision in the right eye was suddenly lost during a fit of sneezing. I saw him five hours after this accident and found a large, fresh hemorrhage covering the macula. This new extravasation contained a round, sharply defined, brilliantly-white center.

The stereogram presents the ophthalmoscopic picture seen at this time. Although the patient has a very dark complexion, the fundus possesses a light, yellowish cast and the surface appears granular. This appearance is attributed to an orange colored choroid shining through deeply pigmented retinal epithelium. The optic disc has a uniform pink color, but there is no evidence of neuritis. The axial light streak on the arteries is unduly broad and bright. The veins are rather full. The entire fundus contains numerous striate hemorrhages several of which contain a round, white, central spot. Immediately below an artery, on the nasal side, is a large preretinal hemorrhage. Beneath the supranasal vessels is a yellowish patch, about the area of the optic disc, which marks the site of an old retinal hemorrhage.

The patient returned to Syria where he died from leukemia about one year later.

**RETINITIS IN PERNICIOUS ANEMIA.** The retinitis of pernicious anemia is largely hemorrhagic in type. The ophthalmoscopic appearances resemble those of hemorrhagic leukemic retinitis, although Schöbl sees certain peculiarities in that of pernicious anemia, *i. e.*, a greyish cast to the fundus, dark veins and nearly empty arteries. Müller declares that retinal hemorrhage occurs in every case of pernicious anemia when the disease is at its

height. Certainly, it usually has been found when sought. This point is of diagnostic importance as retinal bleeding is very rare in chlorosis and simple anemia. The hemorrhages vary in size and occur in all layers of the retina. White areas appear, said to be identical with those found in leukemia, but in reported cases no distinction is made between the spots in new and old extravasations. Microscopic examinations have shown that the spots in pernicious anemia contain lymphocytes, phagocytes and cytoid bodies (ganglionic nerve fibers). Apparently these spots vary little in structure from those found in any old retinal hemorrhage. The extravasations are said to be especially frequent around the nerve.\* Retinal edema, white and yellow spots in the retina and a star figure at the macula have all been seen in pernicious anemia.† Choked disc is not an unusual complication. Many authors have mentioned the presence of vascular degeneration in this disease and both Moses and Michel have met thrombosis of the central vein.

### *Bibliography.*

- Amman, Beitr. z. p. A. XXXVIII, p. 1.  
 Bettmann, A. f. A., XI, 1, p. 23.  
 Bull, C. S. Am. J. Med. Sci., 1879; Trans. Am. Oph. Soc., 1886.  
 De Schweinitz, in Tyson's "Brights and Diabetes," Trans. Am. Oph. Soc., 1896, 645.  
 Dodd, A. f. A., XXXI, 1895.  
 Galezowski, Recueil d'O. 1873, p. 90.  
 Ginsberg, Arch. f. Oph. LXXXII, 1.  
 Greenow, Parsons Path. of The Eye, IV, p. 1297.  
 Hausen, Nord. Med. Ark., XII, 1, 1880.  
 Hawthorne, Lancet, Sept. 30, 1899.  
 Hirschberg, C. f. p. A., X, 1886; XV, 1891, p. 18.  
 Hirschmann, Inaug. Dissert., Berlin, 1886.  
 Howe, Am. J. of Oph., 2, 1885, p. 116.  
 Immermann, Deutsch. Zeitschf. f. Klin. Med. XIII, p. 209.  
 Knapp, A. f. A., X, 1, 1880, p. 99.  
 Kramszky, Jahresbericht f. Oph. 1878, 226.  
 Leber, Graefe u. Saemisch Handb. Bd. 5. Zeits. f. Med. Blätter, Bd. VII, 1869.  
 Litzmann, Deutsch. Klinik, 1852.  
 Magnus, Ref. in Norris & Oliver, III, p. 517.  
 Mauthner, Wein Med. Klin. Rundschau, 1893.  
 Michel, Deutsch. Arch. f. Klin. Med., XXII, 5 u. 6, 439; 2. Auf. Lehrb.  
 Moses, Inaug. Dissert. Wurtzburg, 1896.  
 Müller, Die. Prog. Pern. Anämie, Zurich, 1877; Ref. in Jahrb. f. O. 1877, 207.  
 Nettleship, O. H. Rep., XV, 4, 1903; XVI, 1, 1904; XI, 1886.  
 Parsons, T. O. Soc., XXIX, 1909, p. 160; XXVII, 1907, p. 119-120; Path. of the Eye, p. 1294.  
 Possauer, Inaug. Dissert. Zurich, 1894.  
 Quincke, Deutsch. A. f. Klin. Med., XX, 1; Volkmann's Samm. 1876.  
 Roth-Schirmer, A. f. Path. Anat. XL, p. 441.

\*Uthoff; Amman.

†Immermann; Bettman; De Schweinitz; Sgrosso; Müller; Hausen; Quincke.

- Saemisch, *Zeitsch. f. Med. Blätter*, Bd. VII, 1869.  
 Schöbl, Norris and Oliver, III, pp. 505-512-522.  
 Schlesinger, *Inaug. Dissert.* Berlin, 1884; Ref. in Norris & O., III, p. 523.  
 Sgroso, ref. in *Jahresber. f. Oph.* 1898, p. 495.  
 Silex, *Berlin Klin. Woch.* 1895, p. 385.  
 Simon, C. f. p. A., 1894, p. 132.  
 Steffan, *Jahresbericht f. Oph.* 1873, p. 348.  
 Uthoff, *Intern. Med. Cong.*, Paris, 1900; *Klin. m. f. Aug.* 1880, p. 513.  
 Wilbrand u. Saenger, *Path. d. Netzhaut*, p. 316.

**ANGIOPATHIC SYPHILITIC RETINITIS.** In the eye syphilis exhibits a preference for the uveal tract and when it invades the fundus it usually appears as a chorioretinitis (Stergms. 19-20-21). Exceptionally, the brunt of the disease is borne by the retina, and in rare cases only the retinal vessels are affected. Syphilitic disease limited to the inner retinal layers was first described by Jacobson and later by Mauthner. The existence of such a condition has long been contested. Schöbl described five cases which he considers typical of the condition. The symptoms were: dusty opacities in the posterior part of the vitreous; red blurred disc; slight retinal edema; arteries narrowed; veins dilated; retinal hemorrhages. The presence of vitreous opacities in Schöbl's cases would indicate that the choroid as well as the retina was affected. A review of the literature demonstrates that most cases present the retinal changes incident to degeneration of its vessels, differing in no way from angiopathic retinitis as it occurs in other diseases. A wall-like opacity about the nerve similar to that which occurs in albuminuria, was found in syphilitic retinitis by Classen, Schweigger and Alexander. Groups of minute, yellowish dots, clustering about the blood vessels were described by Hirschberg. An ophthalmoscopic picture identical with that seen in albuminuric retinitis has been observed in syphilitics whose urine contained neither albumin nor sugar, by Knapp, Haab and Schaffels. Arteriosclerotic alteration in the retinal and choroidal (Fig. 80) vessels without marked changes in the retina is a matter of common observation in cases of old syphilis. Haab regards white scales on the retinal vessels as characteristic of syphilitic vasculitis. Cerebral syphilis frequently is accompanied by sclerosis of retinal vessels. v. Hösslin describes five cases of brain syphilis which exhibited endarteritis of the retinal vessels, and in one of the cases there was optic neuritis. The term "hemorrhagic syphilitic retinitis" has been applied to cases in which syphilitic vasculitis resulted in extensive retinal hemorrhages. Closure of the central vessels and their branches occurs in syphilis as in the vascular degeneration of other diseases. Closure of the central artery has been described by Stölting and by Oglesby, and Haab regards syphilitic endarteritis as responsible for many

**cases.** Glaucoma developing in syphilitics has been attributed to specific vascular degeneration.\* Before accepting this conclusion it is necessary to demonstrate that glaucoma can be directly produced by disease of the blood vessels.

Visual disturbance in syphilitic retinitis may be out of proportion to the visible changes. Glimmering is an early and prominent symptom which Hirschberg attributes to retinal anemia caused by arterial disease. It is, however, more reasonable to suppose that "glimmering" is caused in these, as in other cases, by choroidal disease. The same explanation applies to night blindness, which is a frequent symptom of syphilitic retinitis. Ring scotomata and other forms may develop. In syphilitics with angiopathic retinitis, the urine may contain albumin or sugar. In such cases the retinitis should not be classed as strictly syphilitic.

### *Bibliography.*

- Alexander, Syph. u. Auge, 1895, 22; 67 Versam., d. Nat. u. Artz z. Lübeck, 1895, 192.  
 Classen, A. f. O., X, 2, 137.  
 Galezowski, Annal. d'Ocul., CXIV, 389.  
 Haab, Atlas u. Grund. d. Ophthal. 888 auf. Fig. 369; Festschrift f. Helmholtz 1891, Norris & Oliver, IV, 528.  
 Heubner, D. Syph. Gefässerkrank. d. Gehirnarterien, Leipzig, 1874.  
 Hirschberg, Klin. Beobach, 1874; Beitr. z. A., 1, 1876, und 3, 1878; Berlin. Klin. Woch. 1888.  
 Von Hösslin, A. f. Klin. Med., LXVI.  
 Jacobson, Konisberger, Med. Jahrb. Bd. 1, H. 3, 283; Bezeih. d. Verand. u. Krank d. Sehorgans z. Allgemeinleiden, 24.  
 Knapp, A. f. A., IV, 205.  
 Mauthner, Lehrb. d. Ophthalmoscopie, 370.  
 Oglesby, Willbrand u. Saenger, Die Path. d. Netz., IV, 1, p. 352.  
 Panas, A. d'O. XXII, 69.  
 Samelsohn, Soc. franc. d'Oph., 1891.  
 Scheffels, Deutsch. Med. Woch., 1897.  
 Schöhl, Norris & Oliver, III, 480.  
 Schweigger, Vor. u. d. Gebrauch d. Augenspiegels, 110.  
 Stölting, A. f. O., XLIII, 306.

**RETINITIS CIRCINATA.** Fuchs, in 1893, reported a series of twelve cases, presenting a uniformity of symptoms that entitled them to be classed as an independent affection which he designated as "retinitis circinata." The condition was not unknown previous to Fuch's paper, Hutchinson having first described it, in 1876, as "a symmetrical, central, chorio-retinal disease, occurring in senile persons." Subsequently a number of

\*Panas; Samelsohn; Alexander; Galezowski.

cases were reported as "Hutchinson's changes" and under other names.\* The prominent feature from which retinitis circinata derives its name is a girdle of white spots encircling the macula. The characteristic changes usually are confined within the triangular space enclosed between the superior and inferior temporal vessels. In typical cases, the macula around the fovea presents a greyish opacity. At a certain distance this opacity is surrounded by a ring of large and small, milk-white spots. Between the ring and macular opacity the retina may or may not appear normal. Generally, the girdle of spots is horizontally elliptical in outline and incomplete on the nasal or temporal sides. The periphery of the girdle usually falls short of the larger temporal vessels and the disc, but in certain cases both may be included within its radius. The vessels pass over the spots when the two are in contact. The spots are not bordered by pigment. When first formed they are round, but may coalesce and produce lobulated forms or even a white band with scalloped edges. The spots give the observer an impression that they are slightly elevated. The diameter of the macular opacity always is greater than that of the optic disc, and in fresh cases it may appear sharply defined. Later, the opaque area may be extended and swelling of the retina is demonstrable. Several cases have been reported in which the retina was detached in the macular region. Pigmentation does not belong to the ophthalmoscopic picture, but stippling and pigment spots have been observed outside the girdle. Within the girdle, a delicate haze hangs over the zone of apparently normal retina. In old cases, hemorrhages occur in the affected region, usually along the vessels. According to Fuchs, bleeding occurs in about half of the cases. Holms Spicer found hemorrhages in the fellow eye. Fresh spots and new hemorrhages may form. In very old cases pigmentation may occur. The optic disc is unaltered unless touched by the girdle, in which case its outline may be blurred. The retinal veins are tortuous and the vessels may present other arteriosclerotic changes. In course of time the retina included within the girdle atrophies. The spots may long remain unchanged or may break up. Fuchs has seen the macular opacity and spots disappear in four years and in another case continue for seven years. DeWecker has observed a case for twenty years. The spots sometimes change color and exhibit a dirty-grey stippling, and cholesterine deposits appear. In some cases a proliferative process ensues and the retina becomes greatly thickened from formation of connective tissue.

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\*Galezowski in 1886; Goldzieher in 1887; deWecker and Masselon in 1891 as *dégénérescence graisseuse*; deWecker in 1894 as *dégénérescence blanche*.

*Retinitis circinata* is a rare disease. Among 70,000 eye patients, Fuchs found eight typical and four atypical cases. DeWecker saw fifteen cases among 150,000 patients. In Fuch's twelve cases, only one eye was affected in seven and both eyes in five. It appears to be more common among women than among men. Generally the subjects are over 60 years of age and have arteriosclerosis; cases occur, however, at all ages. DeWecker saw a case in an infant. Great visual disturbance always exists, but develops gradually. It consists of a relative central scotoma, with a diameter of 10-12 degrees, which soon becomes absolute. Peripheral vision is not affected, but old cases develop concentric contraction of the field. The light sense remains good. The patients do not complain of night blindness, day blindness or metamorphopsia, from which it is inferred that the rod and cone layer of the retina is unaffected. The course of the disease is chronic and progressive and the prognosis is bad, but absolute blindness does not result, peripheral vision being retained.

The nature of the process and morphology of the spots have not been determined. Fuchs regards the latter as exudates similar to those found in albuminuric retinitis and denies that they are connected with hemorrhages. Goldzieher thinks the condition is a manifestation of arteriosclerosis, the spots being necrotic areas caused by closure of arterioles. Nuel accepts arteriosclerosis as the cause and believes the spots to be in Henle's layer which is best developed in this region. DeWecker declares that the spots are blood clots which have undergone fatty degeneration. Only one case, that of Ammann, has been microscopically examined. Unfortunately, the authenticity of the case may be questioned, as the patient had advanced glaucoma and albuminuria. In the affected part, the retina had four times its natural thickness, due to thickening of the internuclear layer. Ammann decided that the spots were clusters of fat granules and hyaline masses resulting from disintegration of blood; but the blood had all disappeared. Ammann's conclusions are not final. Clinically, bleeding is an inconspicuous feature in *retinitis circinata*. When it occurs it may or may not leave a white spot, but there is no doubt that the characteristic garland of spots occurs independently of hemorrhages. The arrangement of the spots around the macula corresponds to the terminals of the perimacular arteries, which strongly suggests nutritive disturbance from vascular disease. The gradual development and progressive character of the affection indicate a chronic degenerative process which, from its situation, may be regarded as of vascular origin.



**STEREOGRAM 40. RETINITIS CIRCINATA.\*** Right eyeground of woman 60 years of age. In her first confinement, 31 years ago, she had puerperal eclampsia. Eight subsequent confinements were normal and all children born are still living and healthy. Urine normal. Blood pressure 180-190 mm. Hg. Vision of the right eye has been progressively failing for the past year. Aside from slight vascular changes in the retina, the left eye appears normal. In the fundus of the right eye is an open, white girdle around the macular region. The radius of the girdle fills the space between the superior and inferior temporal vessels. The gap in this circle occurs on the side nearest the nerve, and is filled by a yellowish-white strip resembling an infiltration of the retina. The girdle is composed of large and small, closely packed, white spots, sparsely stippled by bluish specks resembling slight surface depressions. Many of the spots have coalesced, but between most of them a line of demarcation can be traced. Numerous small, round, white dots border the spots and coalesce with them. Above and on the outer side of the girdle are numerous small, white and greyish spots with diameters approximating that of the central vein. Below, in the radius of the white circle, is a mass of branching pigment, lying upon a patch of discolored retina. Adjoining, are a few similar but smaller spots. Within the girdle the entire retina is opaque and appears covered by a grey haze. The optic disc contains a deep physiologic excavation and appears normal. The retinal arteries present bright central light streaks. The terminal of the superior temporal artery runs in front of the white girdle and in this part of its course is dilated and tortuous. The retinal veins are slightly dilated. Two striate hemorrhages are seen along the course of the inferior temporal vein, immediately outside the girdle.

**DIAGNOSIS.** It is not difficult to diagnose retinitis circinata if attention is once directed to the characteristic arrangement of the white spots. In retinitis circinata white spots never appear in the macula, but in albuminuria and diabetes the macula is the favorite site of exudation.

#### *Bibliography.*

- Amman, A. f. A. XXXV, 1897; A. of Oph., XXVII, 1898, 203.  
 DeWecker, a. d'Oph. clinique, Paris, 1886.  
 DeWecker-Masselon, Oph. clinique, Paris, 1891.  
 Fuchs, A. f. O. XXXIX, 3, 1893.  
 Galezowski, Traité icon. d'Ophthalmoscopie, III, Paris, 1886.  
 Goldzieher, Weiner Med. Woch. 1887; B. d. O. G., 1896; A. f. A. XXXIV, 1897.  
 Hutchinson, O. H. Rep., VIII, 1876.  
 Nuel, A. d'Oph., XVI, 1896.  
 Spicer, T. O. Soc., XIV, 1894; XVI, 1896.

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\*I am indebted to Dr. Ward S. Holden for the privilege of reporting this case.

## CHAPTER VII.

### CLOSURE OF THE RETINAL VESSELS.

**CLOSURE OF THE CENTRAL ARTERY (Embolism).** For the purpose of practical discussion, the retinal circulation may be regarded as a terminal system. Such collateral vessels as exist are insufficient in size to maintain the vitality of the tissues; consequently, occlusion of the lumen in the central artery is immediately followed by acute retinal anemia and loss of vision.

**OPHTHALMOSCOPIC APPEARANCES.** If viewed immediately after the circulation has been interrupted, the following characteristic picture is presented. The media are clear; the fundus has a dull, slight grey cast; the optic disc is pallid; the arteries are reduced in size and the smaller twigs are indistinguishable. Frequently the arteries appear fairly well filled at the periphery and filliform at the disc. If arteriosclerosis has rendered the arterial walls opaque, they appear as white cords or show irregular constrictions with apparent interruptions of the blood column. As a rule, the veins can be traced throughout their courses, but frequently their proportions appear reversed, *i. e.*, they are widest at the periphery of the field and diminish in size as the disc is approached.

After closure of the central artery degenerative changes in the retina immediately ensue. Within a short time a white, cloudy, edematous-looking opacity appears at the posterior pole and gradually extends over the fundus. The fovea centralis, however, retains its normal color, appearing as a "cherry-red" spot, strongly contrasting with the surrounding white. The margin of the disc becomes indistinct and surrounded by delicate, radiate striations, which are never pronounced as is the case in tumescence of the nerve head. The retinal opacity reveals small vessels around the macula which ordinarily are invisible. At the periphery, where the retina is thinnest, the opacity gradually disappears. Retinal hemorrhages, usually insignificant in size, are sometimes present around the disc and macula. After a variable time (hours or days) the retinal circulation is, in a measure, re-established. When this first takes place, a remarkable phenomenon, known as the granular current, may occasionally be observed. In this condition the blood columns of certain vessels, more commonly the veins,

are broken, presenting the appearance of moving red beads separated by clear interspaces. The rapidity of the current varies and abrupt fluctuations may take place. Thus, the blood stream may be moving steadily through a vessel when, suddenly, it becomes slower or even reversed; or a to-and-fro (pendulum) movement may occur. Rarely all motion ceases. Pulsatory entrance of blood into the vessel is very rare. I have seen but one case. After circulation is partially restored, if the arteries are not sclerosed, the slightest pressure reproduces the granular current. Atrophy now ensues. The opacity disappears and the retina regains its transparency. The outline of the disc becomes sharp and the nerve atrophies. The vessels undergo degeneration, which in some cases is extreme (Stergm. 44). A faint haze always remains over the fundus in the retinal atrophy following closure of the central artery.

STEREOGRAM 41.\* CLOSURE OF THE CENTRAL RETINAL ARTERY (Embolism?). Left fundus oculi of a girl 18 years of age; telephone operator. The left eye has been subject to transitory obscurations of vision. Suddenly, while working, she suffered from slight vertigo and the left eye became blind, but on this occasion vision did not return. The eye was without light perception then or afterward. Urine normal; blood pressure 125 mm. Hg.; no syphilis; no tuberculosis. An aortic systolic murmur was heard by some and not by others.

The stereogram was painted seven days after closure. The pale optic disc is surrounded and obscured by a broad halo of opaque retina. The retina is opaque also in the macular region, strongly contrasting with the "cherry-red" fovea. The larger arteries are filiform and the smaller branches invisible. A few small hemorrhages have occurred around the macula and nerve. The veins are better filled than the arteries. The supranasal vein is broader at the periphery than at the center. Slight pressure on the eyeball produces pulsation in the vessels, proving that circulation is partly re-established.

Two years later the retina was highly atrophic (Stergm. 44). Physical examination at this time failed to reveal any cause for closure of the artery.

INTERPRETATION OF THE OPHTHALMOSCOPIC PICTURE. RETINAL EDEMA. The intraocular distribution of the central artery is limited to the inner layers of the retina; consequently, occlusion of the artery interrupts metabolism in these layers. The ganglion cells with their axons

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\*Patient of Dr. J. H. Ohly, Brooklyn Eye and Ear Hospital.

(the nerve fibers) undergo coagulation necrosis, whereby their transparency is lost. In addition there is more or less serous exudation, analogous to the postanemic edema occurring in white infarction of the kidney, spleen and other organs. Ordinarily, the amount of edema is slight; exceptionally, however, it is abundant (Stergm. 42). Retinal opacity usually appears within two hours after closure, rarely later than the second day. De Schweinitz mentions a case in which he saw it 20 minutes after an attack. Von Graefe's case was remarkable in that it did not appear for two weeks. Schweigger regards early retinal opacity as evidence that the arterial obstruction is nonembolic. It is most marked around the disc where the nerve fiber layer is thickest and around the fovea where the ganglion cells are most numerous. In course of time the waste products of retinal degeneration are removed, after which the inner layers of the retina are converted into a thin cicatrix. The external layers of the retina are nourished by the choroid and therefore are not affected. The supporting tissue of the retina, which is common to all layers, also escapes destruction.

STEREOGRAM 42.\* CLOSURE OF CENTRAL RETINAL ARTERY. Right fundus oculi of a woman, 37 years of age. Urine normal. No trace of syphilis. Heart slightly dilated, but no murmurs. Two weeks ago she suddenly became blind in the right eye, since which time there has been no perception of light. The subsequent history of the case is unknown. Ophthalmoscopic examination revealed the fundus picture of recent closure of the central artery. The retina is opaque throughout the entire posterior polar zone. Around the nerve and at the macula the opacity is very dense, completely obscuring the disc, while the "cherry spot" at the macula is barely distinguishable. That this retina is edematous as well as opaque is shown by opacities which overlie the vessels. Arteries are indistinguishable from veins. Most of the vessels taper from the periphery toward the disc. An ascending and descending vessel appear as white lines without visible blood columns, indicating that their walls are sclerosed.

FOVEAL RED SPOT. The ganglion cells and nerve fibers of the retina do not extend over the fovea centralis; therefore, its normal red color is not obscured by the retinal opacity which appears after closure of the central artery, on the contrary, the fovea becomes highly conspicuous by contrast with the surrounding opaque ganglion cells. The preceding explanation of the foveal red spot was given by Von Graefe and may be accepted as true. Other less satisfactory theories have been advanced;

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\*From the service of Dr. J. Scott Wood, Brooklyn Eye and Ear Hospital.

*i. e.*, hemorrhage (Blessig, Steffan); circumscribed chorioretinitis (Nettleship); retinal pigment (Fischer); attenuation of retinal elements and atrophy of pigment (Elschnig); hyperemia of the choroid (Loring). Of these theories, only hemorrhage has received any support. Hemorrhages near the macula frequently assume a rounded form and are easily mistaken for the red fovea (Stergm. 48). Very rarely, however, bleeding does occur in the fovea (Fuchs) in which case it may be regarded as accidental. The cherry spot appears and disappears with the retinal opacity, which would not be the case were it a blood clot. Loring's theory is supported by the case reported on page 113 in which I found great local congestion of the choroid behind the macula. The shape of the cherry spot usually is described as elliptical. Ophthalmoscopically, the apparent form of the spot is modified by position of the light and by shadows cast upon the walls of the macular depression. Anatomically, the fovea is not oval, but round.\* The distinctness of the spot is modified by the degree of opacity with which it is contrasted and also by the amount of retinal edema. If the retinal epithelium or choroid is deeply pigmented, the spot will be darker than in a blond fundus. Inoyu has seen it coal-black in a Japanese.

**RETINAL VESSELS.** Eyes removed during life or after death exhibit arteries diminished in size but still containing blood in which the red cells have agglutinated and separated from the plasma, so that the vessels contain alternate sections of plasma and massed red cells. A similar condition exists in the retinal vessels when the supply of blood is withheld from closure of the central artery. Those vessels which appear empty are, in reality, filled with plasma. Therefore, it is inaccurate to describe an artery in the living eye as empty unless the lumen is abolished by collapse or thickening of its walls. Arteries filled with plasma preserve their contour, appearing, when viewed with the ophthalmoscope, as dull, yellowish-tinted *cords*, differing in this respect from an artery emptied by external pressure with the finger, in which case the vessel appears as a bright, flat *band*. They differ also from highly sclerosed vessels, which resemble white pipe stems. In cases of central artery obstruction, if the retinal arteries are not diminished in size, closure is incomplete. It is, however, difficult to estimate a moderate diminution in the width of the retinal vessels. Mistakes in this respect are frequently made and are best avoided by comparisons with the fellow eye. The arteries adapt themselves to a diminished blood column by elastic contraction of their walls (Elschnig),

\*Piersol; Schmidt-Rimpler; Merkle; Dogiel.

and also as a result of intraocular pressure (Fischer). Unless circulation is quickly restored, the vascular contraction becomes permanent from proliferation of connective tissue and degenerative changes in the walls.

Frequently, the arteries appear better filled at the periphery than on the disc. Elschmig thinks this is due to the greater musculature of the larger arteries which enables them to expel blood more readily than smaller vessels. Reimer, however, states that the musculature of a vessel is proportionate to its size and sufficient to maintain its tone; he regards arteriosclerosis as a more probable cause of the diminished size of the larger vessels. Apparently, a more reasonable explanation than either of the above may be found in the capillary action exerted by the exceedingly small retinal capillaries which, in the *absence of a cardiac impulse*, would hold the blood like a sponge; as the total amount of blood in the retina after closure of the central artery is insufficient to fill all its vessels, it accumulates in the capillaries and the scarcity is manifested chiefly in the larger vessels.\* Although the veins are better filled than the arteries, they frequently are described as tapering towards the disc. This also can be explained by scarcity of blood in the retina and its tendency to remain in the capillaries, in the absence of an efficient *vis-a-tergo*.

In some cases of otherwise complete blindness from closure of the central artery, an area of light perception is retained for a time on the temporal side of the field. Fischer demonstrated that this corresponds to a narrow zone around the optic nerve, which is occupied by capillary anastomoses between the blood vessels of the retina and choroid. This area also becomes blind in the atrophic stage.

In some eyes the macular region is supplied by a so-called cilio-retinal artery in which blood continues to circulate even though the main artery is closed. When this condition exists, central vision alone is preserved. Wilbrand describes a typical case in which, fifteen years after closure of the central artery, a narrow strip of retina, corresponding to the distribution of two cilio-retinal vessels, had retained both normal vision and appearance notwithstanding all other parts of the retina had undergone extreme atrophy. Cilio-retinal vessels are quite common and are recognized by the manner in which they hook over the edge of the optic disc, usually on the temporal side (Stergm. 4). They are supposed to spring from the choroidal system of vessels which would render them independent

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\*In retinitis pigmentosa, optic atrophy and other chronic degenerations of the retina, in which the cardiac impulse is unimpaired, there is comparatively uniform narrowing of the retinal vessels.

of circulatory disturbances in the central artery. It is a question, however, whether they really originate in the choroidal system or are branches from the central artery, given off far back in the nerve, proximal to the usual site of obstruction.

Variations in the appearance of the vessels are occasionally noted in central artery obstruction. Thus, Priestly Smith described a case in which the veins tapered towards the disc but abruptly enlarged at the junction of tributary veins, then again diminished in size until joined by another tributary. A similar condition is shown in stereogram 43. Nettleship describes an unusual appearance, seen thirty hours after closure. At the macula the vessels were bordered by clear bands, wider than the vessels, which were compared to "rills of water in a dried up brook channel." They were too wide for degenerated vessel walls. Reimer mentions a similar appearance in one of his cases.

When the vessels begin to refill it often is impossible to distinguish veins from arteries, both being very dark from deficient oxidation of the blood incident to retardation of the current. Difficulty in differentiation is further increased by the frequent absence of light streaks on the vessels. In the atrophic stage the arteries become more attenuated than the veins (Stergm. 44).

When obstruction of the central artery is at its height it is probable that a meager circulation, derived from collaterals around the nerve head, still exists in the retina. It has been held that inability to produce pulsation in the retinal vessels by external pressure indicates cessation of the circulation. This test is of little value, inasmuch as pulsation is not readily produced in sclerosed vessels, which is the usual condition in closure of the central artery.

Clinically, when the retinal circulation is very feeble, it is completely arrested by the slightest pressure. Reimer asserts that a certain amount of circulation is present so long as the blood column remains unbroken, and complete interruption can be assumed only when the blood column is broken and the separated portions stand still. This statement is founded upon the ophthalmoscopic appearance of the retinal vessels at death. It must be remembered that the appearance of a broken blood column may be simulated by opaque spots in the walls of a vessel, which obscure a thin but continuous column of blood. If either spontaneous or pressure pulsation can be demonstrated it is, of course, positive proof that circulation exists. The retinal circulation will be restored, sooner or later, after its apparent suspension. At this time the arteries may only partly

regain their former calibre or, on the other hand, become hyperdistended. In the latter case small hemorrhages occur (Haab).

If blood is absolutely withheld from the retina, the ganglion cells perish in a very short time. Under similar conditions in the brain the cells die within half an hour or less. The clinical histories of cases in which vision has been completely or relatively restored several days after obstruction of the central artery, indicate that sufficient blood was supplied to the retina to preserve tissue vitality, although insufficient to maintain functional activity.\* If visible degenerative changes in a retinal artery have produced occlusion, there will be blindness throughout the area of its distribution.

**GRANULAR CURRENT.** When the blood current in a vessel is sufficiently retarded, the red corpuscles agglutinate into masses which float in the blood plasma. The size and appearance of these corpuscular masses vary with the rapidity of the current. Thus, if the blood stream is comparatively swift, they are small, resembling rapidly moving grains of sand, while in a sluggish current the corpuscles form long, dark red cylinders, separated from each other by clear spaces occupied by plasma. The ends of the cylinders present a sharply indented boundary line like the fractured end of a cast iron bar (Reimer). A similar separation of the corpuscles from the plasma is seen in the vessels of enucleated eyes. The granular current appears first in the veins, where the blood stream is slower than in the arteries. If a granular current appears in an artery, while the corresponding vein presents an unbroken blood column, the vein receives blood from areas additional to that supplied by the artery. Cessation or reversal of a blood current in a vessel may be due to intermittent blood supply or, according to Elschmig, to influx of venous blood from a region of higher arterial pressure. The amount of fresh blood supplied by a granular current must be insignificant. In one of my own cases (Stergm. 43) it barely sufficed to preserve light perception, although, as shown by Raehlman in a case of extreme arteriosclerosis of the retinal vessels, only a small quantity of blood is required to maintain the function of the retina.

**HEMORRHAGE.** In closure of the central artery extravasations of blood are few and insignificant. In 155 cases of trunk closure collected by Fischer, hemorrhage occurred in 47. The extravasations are feathery in type and usually appear in the areas of capillary anastomoses around the disc and macula when the circulation is first restored. Haab says that if

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\*Hirschberg, Noyes, Alexander, Hasse, Perles and others.



the vessels are hyperdistended, bleeding occurs, but it is uncommon when the vessels are incompletely refilled. These observations apply to secondary hemorrhages which usually appear in from two to twelve days after arterial closure and are coincident with refilling of the retinal vessels.

Thus far, in this discussion, it has been assumed that acute retinal ischaemia (white infarction) always follows closure of the central artery. While this is true for the great majority of cases, it is possible that under certain conditions obstruction in the central artery may be followed by the picture of hemorrhagic retinitis (thrombosis). I know of no case in which hemorrhagic retinitis has been preceded by the ophthalmoscopic picture of central artery closure; therefore, if hemorrhagic retinitis is ever caused by obstruction in the central artery, the bleeding occurs at the onset of the attack; in other words, it is a case of hemorrhagic infarction. That such an event ever occurs is doubted by Michel, Haab, Coats and Harms. Coats and Harms accord in the statement that, in every case presenting the appearance of retinal thrombosis, the central vein will be found obstructed if it is properly examined microscopically throughout its entire course. Nevertheless, certain cases, particularly one by Reimer, appear to prove that, under certain conditions, hemorrhagic retinitis is due to obstruction in the central artery. In denying the possibility of such an event, writers appear to be unfamiliar with the experiments of Wm. H. Welch, who conclusively proved that, in a terminal circulation, hemorrhagic infarction invariably occurs when the trunk artery is *partially obstructed* and the blood pressure on the distal side in front of the obstruction, falls to  $\frac{1}{4}$  or  $\frac{1}{5}$  of normal. If, however, the artery is suddenly and completely closed, there is no bleeding (Stergm. 42). It must be remembered that very few cases of either central artery or central vein closure have been microscopically examined by competent pathologists; therefore, the following authoritative examination by Reimer is almost conclusive.

The ophthalmoscopic picture of narrowed arteries, tortuous veins and extensive hemorrhage led to the clinical diagnosis of thrombosis of the central vein. Glaucoma developed and the eye was enucleated. Transverse serial sections were made of the posterior part of the globe and 5 mm. of the optic nerve, and every section was examined. The lumen of the central artery was *almost* completely blocked by endarteritis proliferans. The walls of the central vein were thickened but its lumen was free.

I am further strengthened in my conviction that arterial obstruction may produce destructive retinal hemorrhage by the following case:\*

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\*From the service of Dr. Francis Valk, N. Y. Post Graduate Hospital.





Fig. 79.

Fig. 79—Granular Body of unknown origin, containing Nuclear-like Spots, in the Inner Retinal Layers in Albuminuric Retinitis.

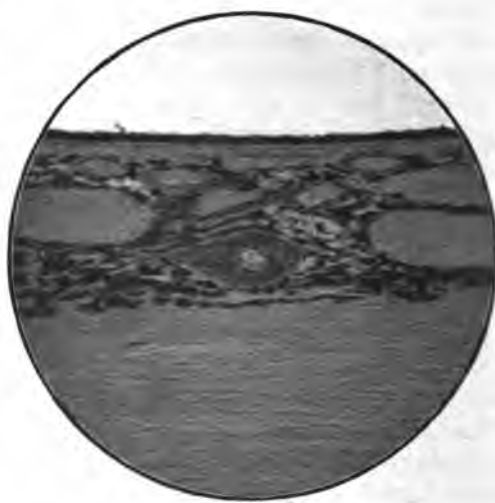


Fig. 80.

Fig. 80—Sclerotic Changes in the Choroidal Vessels in Syphilis, without marked changes in the Retina.

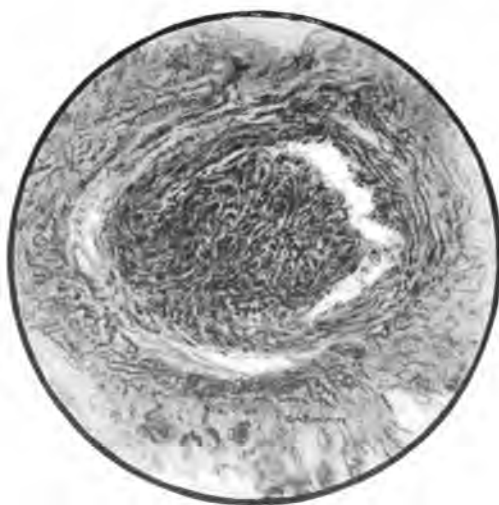


Fig. 81.

Fig. 81—Closure, nearly complete of the Central Artery by Endarteritis Nodosa.

Unmarried woman, 22 years of age. When first examined ophthalmoscopically, the eye presented the picture of hemorrhagic retinitis. Later, intravitreal hemorrhage occurred, glaucoma developed and the eye was enucleated. The nerve was sectioned transversely throughout the entire course of the central vessels. The central vein was not obstructed but the lumen of the artery was *nearly* closed by endarteritis nodosa (Fig. 81).

In both Reimer's case and my own and in Welch's experiments, hemorrhage was due to *incomplete* closure of the artery.

ETIOLOGY. Closure of the central artery usually occurs in the subjects of cardiac or vascular disease, which in many instances, although present and progressive, cannot be demonstrated. The majority of my cases have been young adults in whom arteriosclerosis often is a local disease. In the cases shown by stereogram 41 the only reasonable explanation of the closure was local disease of the artery. In such cases, further manifestations of arteriosclerosis may be delayed for years. In a girl nineteen years of age, observed by Herter, there was sudden closure in the lower main retinal artery. Although apparently in perfect health at the time, edema of the feet appeared within a year. Sometimes the underlying cause is undiscoverable, as in a case cited by Logetschnikow, in which closure of the central artery occurred in a girl of thirteen. The most exhaustive examination revealed no abnormality to explain the closure. Gunn has reported a similar case. As a rule, in closure of the retinal vessels, thorough examination will reveal evidence of vascular disease. In a boy 17 years of age, with closure of the left central artery, Dr. Glentworth Butler found all surface arteries hardened and the blood pressure 150 mm. Hg. There was no cardiac disease and the urine was normal.\* Aside from certain exceptional cases closure of the central retinal artery has been attributed to, (a) arteriosclerotic obstruction; (b) thrombosis; (c) embolism and angiospasm. These causes are supposed to act singly or in combination.

EMBOLISM. In 1854 Jaeger published a typical case of central artery closure, in which he gave the first accurate description of the granular current, but he was unable to explain the cause of the condition. The phenomena were first interpreted by Von Graefe, in 1859, who diagnosed a similar case as embolic occlusion of the central artery. Although received with incredulity, Schweigger confirmed the diagnosis two years later by necropsy. This triumph of V. Graefe's genius was complete and

\*From Brooklyn Eye and Ear Hospital.

to many ophthalmologists "embolism" still suffices to explain any obstruction in the central artery. The chief point of v. Graefe's discovery, namely, that the clinical picture is produced by closure of the central artery of the retina, was admitted, but doubts were soon expressed as to the embolic nature of the obstruction in all cases. That these doubts were well founded has since been demonstrated by microscopic examinations, which show that closure usually is caused by some process other than embolism. At the present time nearly all authoritative writers\* consider that in all cases so far reported as embolic the diagnosis is doubtful. While it is true that an occluding plug has been found in many central arteries, the deductions therefrom must be accepted with reserve, inasmuch as nearly all debatable cases were examined a year or more after closure, at which time it is difficult, if not impossible, to determine whether a fibrinous or calcareous plug was conveyed from a distance or developed *in loco*, the great problem being to separate the secondary histological changes from the primary. Shiba reports that he produced embolic closure of the central artery and choroidal vessels by injecting a quantity of *soft* paraffin into the carotids. Such experiments are useful to demonstrate the tissue changes following arterial occlusion, but prove nothing as to the probability of an embolus blocking the central artery in man.

Anatomical obstacles to the passage of an embolus from the general circulation into the retinal vessels are, the rectangular separation of the ophthalmic artery from the carotid and also of the retinal artery from the ophthalmic. While it may be improbable that an embolus would traverse this route, it is by no means impossible. A more convincing argument against the embolic theory is founded upon certain clinical features of the disorder, such as prodromata; recurrent attacks; occasional involvement of both eyes, or of both central vessels in the same eye; recovery of one eye but not of its fellow; frequent incompleteness of the obstruction and in all cases re-establishment of the circulation, all of which are inconsistent with the presence of an embolus in a terminal artery. Of these objections, discussion has mostly centered around that of restored circulation, and many ingenious theories have been advanced to explain re-

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\*Haab, Harms, Hirschberg, Raehlman, Michel, Wilbrand, Reimer and others. Harms would not diagnosticate embolism unless: (1) The closure occurred from deposit of a body in the lumen of a vessel, which before the obstruction, was everywhere free, and primary disease of the vessel walls could absolutely be excluded; (2) The source of the embolus is found either in valvular disease of the heart or other condition which leads to thrombus formation; (3) The symptoms indicating thrombotic closure are not present.

filling of the retinal vessels a few hours or days after the central artery has been occluded by an embolus. The theory of collateral circulation which has persistently been invoked for this purpose, rests on the following anatomic basis. The arterial circle of Zinn which surrounds the head of the optic nerve, sends branches to both choroid and retina, thus establishing communication between these otherwise independent vascular systems. Unfortunately, however, these collaterals are little more than capillaries engaged in local nutrition and are wholly incapable of suddenly responding to the demand of an ischemic retina for sufficient blood to preserve its vitality. Nettleship has pointed out that the location of a plug in the central artery modifies the amount of blood from these collaterals. Thus, if the artery is closed on the distal side of the lamina cribrosa the vessels from Zinn's circle will be blocked. On the other hand, if the obstruction is situated behind the lamina, these vessels will have full play. This explains certain cases in which, although the artery is closed, the disc retains its rosy hue until the atrophic stage.

Nettleship and Gonin have each reported cases in which a collateral circulation was ophthalmoscopically visible between small vessels from the optic disc and an obstructed retinal artery. The following similar case is the only instance I ever saw of this phenomenon.

STEREOGRAM 43.\* CLOSURE OF CENTRAL RETINAL ARTERY WITH VISIBLE COLLATERAL CIRCULATION. Left eyeground of a woman, 47 years of age, with chronic interstitial nephritis. The right eye presents a typical ophthalmoscopic picture of albuminuric retinitis. She has experienced occasional, transitory obscurations of vision in the left eye. Five hours before coming under observation this eye suddenly became blind. Vision reduced to perception of light. The ophthalmoscopic picture is characteristic of recent occlusion of the central artery.

A typical foveal spot is situated in the center of a discoid area of opaque retina. The optic nerve is well outlined but surrounded by extremely delicate radii, evidently an early stage of opacity. Elsewhere, the retina has a grey, lifeless appearance. A few prominent, white dots, scattered about the macular region, are regarded as products of retinal degeneration due to the nephritis. The visible arteries are highly sclerosed and appear as white cords, with the exception of the supratemporal branch, which contains a sluggish granular current. The corresponding vein also exhibits a granular current, but less broken than in the artery. All veins appear to diminish in size as they approach the disc. The blood

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\*Case was treated by Dr. S. M. Payne, at the Manhattan Eye and Ear Hospital.

visibly circulating in the supratemporal artery is supplied by a plexus of small vessels, situated on and around the margin of the optic disc. The communication between artery and collaterals is plainly seen. On the disc the artery is white and contains no visible blood until joined by the newly-developed vessels, at which point the granular current commences. Pressure on the globe failed to produce pulsation but checked the circulation. Two days later circulation was re-established throughout the retina. The granular current, collateral vessels and macular spot had all disappeared. Those vessels which, during the period of closure, appeared like white cords, now exhibited white bordered blood columns.

There can be little doubt that, in this case, the collateral vessels came from the circle of Zinn or from the disc plexus. The obstruction in the central artery of the left eye was regarded as endarteritis obliterans which had greatly narrowed and occasionally closed the lumen. The gradual advance of such a process combined with occasional interruptions of the circulation, would favor a high development of the collateral vessels. That the collaterals were visible may have been due to unusual size or, as Gonin surmised in his case, they were more superficial than usual. The subsequent restoration of circulation throughout the retina cannot be attributed to the collateral vessels which, although remarkably developed could furnish only an insufficient supply of blood for a single arterial branch. The collateral vessels around the disc may, in time, undergo considerable enlargement but never develop sufficiently to restore circulation in the retina, as shown by the course of events when both retinal and choroidal supplies are severed in opticociliary neurectomy. In these cases the blood enters the choroid through recurrent branches of the anterior ciliary arteries and, possibly, through the vena vorticosæ. The new vessels which then form around the nerve head represent granulation tissue which soon cicatrizes and obliterates all vessels on and around the disc. In closure of the central artery circulation usually is restored through the regular channels. Schlodtmann reported a case in which the optic nerve and a nerve tumor were removed, and on the evening of the same day the retinal vessels were filled with blood. I have seen a somewhat similar case. Of course, in these cases, collateral circulation was established through new formed vessels before the tumor was removed. Coats suggests that circulation may be re-established by enlargement of the vasa vasorum. Admitting that such collateral circulation may be established, the interval between suspension and restoration of circulation in central artery closure is too short for such a system to develop. In the case of







Fig. 82.

Closure of Central Artery with marked Congestion of the Choroid (c) on the Temporal Side, which is the right side in the section.

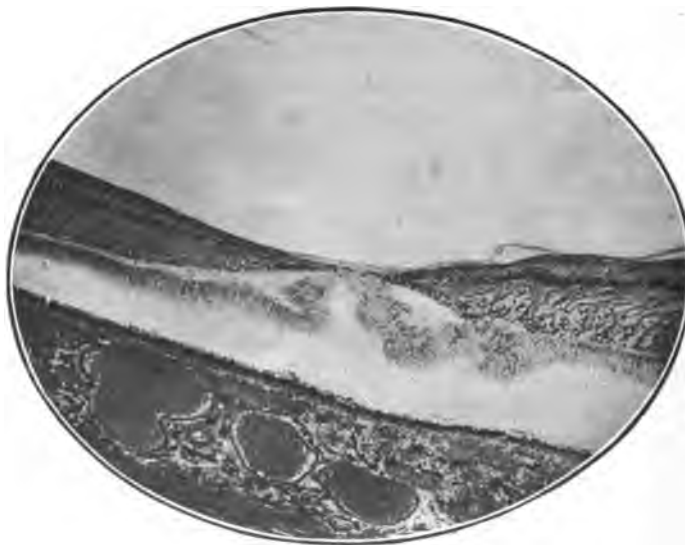


Fig. 83.

The same as Fig. 82, but shows the congestion of the Choroid where it was most pronounced, behind the Macula.

central artery closure described on page 113, there was marked congestion of the choroid on the temporal side, most pronounced behind the macula (Figs. 82-83). The engorgement was purely passive. Is it possible that this local hyperemia was an attempt on the part of the choroid to supply nutriment to a starving retina.\*

A variety of explanations, other than collateral circulation, have been advanced to account for the prompt return of circulation after embolic closure of the central artery. Schneller suggested an embolus in the ophthalmic artery which, after temporarily closing the mouth of the retinal artery, passed onward, permitting the blood again to enter the retina. Mauthner advanced a similar theory. Such an occurrence would be a pathologic curiosity, whereas, something more generally applicable is required. There is a theory of incomplete embolism, due to irregular shape of the plug which permits blood to leak past its edges. Thus, Elschnig describes how an incomplete embolus grazed the arterial walls and provoked a spasm sufficiently strong to grasp and hold the foreign body until it adhered to the vessel. When the spasm relaxed and the artery dilated, the embolus remained attached to one side only, leaving an opening for the passage of blood. Such theories are very properly rejected by most investigators.† It is evident that an embolus will be driven onward until wedged into the walls of the vessel, where any remaining aperture would quickly be closed by swelling of the intima or formation of a thrombus. The objections stated apply also to Wagenman's suggestion that the plug may be reduced in size by inherent shrinking or compression from contraction of the arterial walls. This idea ignores the process of organization and union with the walls of the vessel which occurs in embolism and thrombosis. Fragmentation and distribution of the embolus among the smaller branches of the artery, as suggested by Fischer, is an unusual event in embolism and, as pointed out by Hirschberg, the presence of such fragments should produce some effect, ophthalmoscopic evidence of which is entirely lacking. Canalization of a soft thrombus or embolus would permit the passage of blood. Although this process has been observed in a vein, it has not as yet been demonstrated in an artery. Furthermore, as remarked by Reimer, canalization requires time.

\*Little is known of the mechanism of collateral circulation beyond the fact that ischemic tissue has the power of dilating any vessel that can nourish it. If it be demonstrated that choroidal vessels habitually dilate in closure of the central artery, the impulse to dilation may arise in the supporting framework of the retina which is common to all layers, and is nourished by the choroid. This tissue does not die in closure of the central artery.

†Leber, Sachs, Reimer, Coats.

Elschnig's view that an embolus may be absorbed, leaving no trace of its existence, is unsupported by pathological evidence.

It would appear that each case reported as embolism of the central artery requires the existence of some unusual or improbable event to support the diagnosis. Nevertheless, the possibility that an embolus may lodge in this vessel, as well as the inherent difficulties attending its demonstration, must be admitted. While the clinical aspects of a case cannot be regarded as of much value in differentiating embolism from other conditions, it may be well briefly to recall a historic case. Under the now discarded term of "infiltration of the retina," v. Graefe was familiar with the ophthalmoscopic picture since interpreted as "embolism";\* yet he said of his case that the fundus picture was one he had "never seen before in a case of recent blindness." The optic disc was at no time obscured and the retina remained perfectly transparent for two weeks after the attack. At the end of three weeks, retinal opacity limited to the temporal side had so far developed as to bring out the foveal spot. Schweigger made the microscopic examination and found the central artery blocked by a spherical coagulum at the lamina cribrosa. The chief peculiarity of this case is the long delayed appearance of retinal opacity, a condition which Schweigger considers to be characteristic of embolism. He looks upon the early appearing opacity commonly observed after closure of the artery as an infiltration of the retina produced by gradual slowing of the blood current from endarteritis.

In some cases the artery is closed by a chalky concrement which may be an embolus or, what is more probable, either a thrombus or mass of endarteritic proliferation which has undergone calcareous degeneration. In a case reported by Coats, the eye was enucleated for glaucoma three years after closure of the central artery. The artery was blocked within the lamina by a calcareous mass which had no connection with the walls of the vessel. The plug was considered to be an embolus, a source for which was found in calcareous deposits on the aortic valves. In a case by Manz, the patient died sixteen days after closure. On microscopic examination a structureless mass, possessing the staining qualities of calcareous matter and not connected with the walls of the vessel, was found in the central artery at its bifurcation. In these cases it was assumed that the walls of the arteries were too much degenerated for the foreign body to excite tissue proliferation. In considering these cases it should be borne in mind that manipulations incident to enucleation or preparation

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\*See Schweigger, *Arch. of Oph.* v, XXX, 1901, p. 506.

of a microscopic slide, frequently displace calcareous masses and this sometimes renders it difficult to determine their actual location during life.\*

A source for the embolus is seldom found. Kern analyzed 95 cases presenting the clinical features of embolism of the central artery that were subjected to autopsy. In 66.3% no possible source for an embolus could be discovered, but a diseased condition of the peripheral vessels existed in many of the cases.† Even when heart lesions are found it does not warrant the diagnosis of embolism, inasmuch as heart disease and vascular changes usually coexist. Some authors, while admitting that embolism has been demonstrated, regard it as an exceptional event. Thus, Coats and Parsons see no reason to doubt that embolism occurred in the cases of v. Graefe, Manz, Marple and Coats, while Schweigger regards the case of v. Graefe as the only authentic, recorded case. The majority of authorities, however, await the conclusive demonstration of an embolus in the central artery of the retina.

Usually, an embolus is a detached piece of thrombus. Occasionally it consists of fluid, fat or gas.‡ Theoretically, any of these latter could enter the retina, but no such cases have been demonstrated. Schapringger advanced the view that embolism occurring after a paroxysm of coughing, as in pertussis or after gastric hemorrhage, was due to entrance of gas (air) into ruptured arteries, and he recommended that the patient be subjected to high atmospheric pressure in a pneumatic cabinet as is done in caisson disease. As is well known, incurable blindness may follow hemorrhage from the stomach, bowels or uterus, and, evidently, is not altogether dependent upon the amount of blood lost. Gas embolism offers a satisfactory explanation of such cases.

ANGIOSPASM. When used in this connection, the term spasm should be restricted to a primary contraction of the walls of a blood vessel sufficiently powerful to arrest the circulation, the impulse being imparted through the vasoconstrictor nerves in response to a local or remote irritation. The term should not be applied to the elastic contractility by which the walls of a vessel are passively adapted to a diminished blood

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\*It is conceivable that a calcareous mass may separate from the walls of the central artery and change its position so as to close the lumen. This would constitute embolism.

†In 63 of these cases, 24 had arteriosclerosis, 5 had chronic nephritis and 6 had syphilis.

‡Fat, unless in excessive quantity, is readily dissolved in the blood. Wm. H. Welch believes that, in the living body, gas may be generated in the blood by the *bacillus areogenes capsulatis*.

column. The walls of arteries contain smooth muscle fibers which are innervated by vasoconstrictor and vasodilator nerves. In addition, nerves pass from the periphery to the vasomotor centers which convey centripetal impulses capable of producing the phenomena of reflex irritation. In laboratory experiments the vessels of a part can be emptied or distended by stimulating or inhibiting the vasomotor nerves. On these grounds, neuropathology recognizes a local cerebral anemia due to arterial spasm as causative for certain ailments, such as transitory attacks of aphasia, hemiplegia, monoplegia, hemianopsia, numbness, etc. Among ophthalmologists, spasm was long accepted as a frequent cause for retinal ischemia, particularly for transitory attacks which subsided without permanent injury to vision.\* Recent critical analysis of such cases has thrown great doubt upon this theory; in fact, whenever an opportunity has occurred to make a microscopic examination, some form of actual obstruction has been found, a condition undoubtedly true for nearly all cases of acute retinal ischemia. Occasionally, however, a case occurs in which some other cause is operative. The extreme contraction of the arteries in quinine poisoning is referred to as an example of pure angiospasm, but recent experiments on animals leave little doubt that the contraction is chiefly passive and results from enormous dilatation of the great splanchnic vessels, whereby blood is withdrawn from the peripheral areas† (Parsons). When blood is diverted from the retina in this manner, the ischemia is further increased by intraocular pressure and passive elastic contraction of the arteries. These observations are important as indicating that retinal anemia may be produced also in man independently of any obstruction in the central artery from excessive accumulation of blood in the great abdominal reservoirs. Probably, the retinal ischemia of syncope is produced in this manner. A case in point was reported by Valaude. A man 36 years of age received a blow over the heart which rendered him unconscious for several hours. He presented the ophthalmoscopic picture of bilateral ischemia of the retina. Retinal circulation was not fully restored and atrophy followed.

Hirschberg first described cases in which, after contusion of the bulb, blindness of several minutes duration ensued accompanied by anemia of

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\*In Ole Bull's "Krankheiten der Retinalgefäße." Cramp of the arteries is the dominant pathologic factor throughout the book.

†Vascular degenerations and thrombus formation have been reported as present in the central artery in experimental quinine poisoning (Emil Behse), De Schweinitz confirmed these observations but on re-investigation of his sections he agreed with Holden, that no true endovasculitis or thrombus formation ever occurs in quinine poisoning.

the retina. Pearse has reported such a case following contusion of the cornea. Presumably, there was physical shock with sudden fall of blood pressure, which is an important factor in causing peripheral anemia.\* It is generally conceded that, in traumatic edema of the retina (*Commotio retinæ*) concussion is followed by retinal angiospasm and ischemia which soon is succeeded by vascular dilation and transudation of serum (Stergm. 59). Faravelli tells of a healthy man who suffered attacks of phosphenes followed by temporary loss of vision, first in one eye, later in both. During the seizures the arteries were reduced to threads, while the veins pulsated. After removing a tapeworm, minus the head, the attacks ceased for two months but returned when sections of the worm reappeared in the feces. A complete and permanent cure followed removal of both worm and head. Quaglino describes the case of a literary man who had transitory attacks of blindness attended by sensations of light whenever the meal time was delayed. In the two latter cases an abdominal center of irritation existed, which suggests inhibition of the splanchnic nerves with consequent fall of blood pressure. Ramorino reported two cases of malarial fever in which, during the paroxysm, blindness attended by sensations of light occurred. The arteries appeared filliform and the veins were barely perceptible. In the intervals between the attacks the fundus appeared normal. The cases were cured by quinine. Schnabel describes contraction of the retinal arteries and dilation of the veins during malarial chill. In malarial seizure, the accumulation of blood in the abdominal viscera suffices to explain coincident retinal anemia.

For 23 years I had under observation a patient who had occasional attacks of temporary blindness, during which the retinal vessels were reduced to threads. The first attack followed an almost fatal hemorrhage from placenta previa. At first, both eyes were affected, but during the last ten years of life attacks were confined mostly to the left eye. During the last five years of life arteriosclerotic changes appeared in the left retina. Death resulted from Grave's disease. With the ophthalmoscope, I frequently watched refilling of the retinal vessels. The first attack evidently resulted from exsanguination. The cause of subsequent attacks is a subject for speculation.

Cases of retinal vascular spasm have been attributed to megrim.

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\*Syncope need not, as might be supposed, occur in such cases, inasmuch as diminished blood pressure is felt in the eye sooner than in the cranial cavity, owing to intraocular pressure, which is 20-30 mm. hg., as compared with 10 mm. hg. in the cranial veins. (Hall, experiments on dogs.)

Parisotti describes attacks of ophthalmomiegism during which there was contraction of the arteries in the upper half of the fundus with blindness of the corresponding field. Siegrist found, during an attack of hemicrania, extreme retinal ischemia on the affected side. It may be remarked that glaucoma frequently is mistaken for megrim, and its presence should positively be excluded in making the diagnosis. Ole Bull mistook cause for effect when he said, "I have several times found tension increased while (retinal) arterial spasm continued." The appearance of peristaltic contractions of the retinal vessels, described by Benson, undoubtedly was misinterpreted. Such spasmodic movements in blood vessels is unknown. Cases of sudden blindness, presenting the ophthalmoscopic picture of embolism, have followed subcutaneous injection of paraffin into the nose. These remarkable cases remain unexplained.\* Narrowing of the retinal vessels is said to immediately precede an epileptic seizure.†

Great impetus was given to the doctrine of retinal angiospasm by Raynaud, who gave his name to a group of symptoms consisting of periodic attacks of either pallor or cyanosis, edema, a feeling of impending death and parasthesia of the extremities which frequently became gangrenous. He attributed the condition to spasm of the arteries and, in his enthusiasm, saw isolated segments of the retinal arteries spasmodically contracting. Panas, who ophthalmoscopically examined Raynaud's cases, was unable to see spasm or other circulatory disturbance. Raynaud published his ophthalmoscopic findings as facts. It is needless to say that these observations remain unconfirmed.

Direct, artificial irritation of an artery will induce sufficient spasmodic contraction of the walls to close its lumen; therefore, it is reasonable to believe that the irritating effects of a local arteritis might also excite a reflex angiospasm. Such a case should be classed as one of arteriosclerosis. In view of the uncertainty as to the occurrence of angiospasm in a healthy retinal artery, supposed spasmodic cases should be regarded as secondary to vascular degeneration and the habits of the patient regulated accordingly.

**THROMBOSIS.** Both toxic and bland thrombi occur in the central artery. Toxic thrombus forms in acute orbital inflammation, the walls of the artery being invaded by bacteria. Bland thrombi are due to combined general and local causes. General causes are slowing of the circulation and changes in the blood; local causes are sharp bends in the vessels,

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\*Rohmer; Mintz; Hurd and Holden; Moissioinnier; Silberstein; Uthoff.

†Knies; deBono; Dollo.

narrowing or dilation of the lumen and changes in the endothelial lining. Wilbrand and Saenger cite cases to prove that thrombus may form without coincident arteritis, but Coats, in a critical analysis of the same cases, shows that such an event does not occur. A thrombus formed in another vessel may extend into a normal central artery, as in Siegrist's case where, following ligation of the carotid, a thrombus spread from the point of ligation, through the ophthalmic, into the retinal artery. Analogous cases are reported by Michel, and by Rothmund and Eversbusch. Such cases are unusual and have little bearing on the question as to the frequency with which the clinical picture of central artery closure is due to thrombosis. Reimer regards endarteritis as the sole factor in the production of closure. He argues that in small vessels like the central artery, arteriosclerosis takes the form of endarteritis proliferans, a condition in which, though the lumen be narrowed, the endothelial lining remains smooth and the blood is driven past the obstruction with increased velocity; whereas, thrombus formation requires a slow current and a roughened surface.\* Thrombus formation in an artery is uncommon and usually results from an ineffective arteritis in the course of an acute fever.

Occasional closure of the central artery by combined wall disease and thrombosis appears to be established by microscopic examinations. The following case by Harms must be regarded as authentic.

A man 48 years of age, with hypertrophy of the left ventricle, suffered sudden disturbance of vision in the left eye. After three days the picture of macular artery closure developed with central scotoma. About a month later, rapid obscuration of the entire field occurred and 24 hours later the eye was blind, the ophthalmoscope showing complete closure of the central artery. The eye became glaucomatous and was enucleated. Microscopic examination revealed sclerosis of the central artery which was plugged at the lamina by a well organized thrombus. The macular artery and one other branch were closed by thrombi.†

The slow, progressive manner in which closure took place in this case is considered characteristic of thrombosis.

From the evidence at hand, thrombosis must be regarded as the

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\*Experiments have demonstrated that clotting of blood is accelerated by action of a substance furnished by the walls of the vessels, which has received the name of thrombozym or thrombokinas. It remains to be demonstrated how this coagulating action is modified by mural disease. Zarhelli concludes that thrombokinas is not concerned in thrombus formation, which is a process wholly distinct from clotting. (Bernheim; Zarhelli; Weil; Wolf; Strong.)

†It is always difficult, often impossible, microscopically to differentiate an organized thrombus in the central artery from endarteritic proliferation.



final act in occluding the central artery in certain cases of arteriosclerosis, although it is far less frequent than would be inferred from English and French medical literature.

**ARTERIOSCLEROSIS (Endarteritis Proliferans).** In the preceding pages it has been shown that arteriosclerosis is regarded as the almost constant causative factor in closure of the central artery. Loring, who first advocated this theory, published in 1874 a series of five cases presenting the ophthalmoscopic picture of embolism, each of which manifested some feature which he regarded as inconsistent with the presence of an embolus in the central artery.\*

The mechanism by which endarteritis is supposed to close the central artery may be summarized as follows. At some point, usually the lamina cribrosa, progressive proliferation of the intima diminishes the lumen more and more until only a narrow opening remains. So long as the blood pressure exceeds the elastic contractile power of the artery, the blood will continue to flow past the obstruction; but should the blood pressure sink sufficiently from any cause, such as syncope, hemorrhage, determination of blood to other parts, sleep, etc., or if the opening is closed from swelling or spasmodic contraction of the walls, the heart no longer

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\*The conception that closure of the central artery might be due to vascular disease has, wrongly, been credited to Mauthner. Mauthner advanced no new theory to explain closure; he simply doubted that it was always embolic. He says, "In the majority of cases the pathologic appearances may, perhaps, be ascribed to other affections *as yet unknown*." In a transitory case he suggested that return of circulation might be due to fragmentation and distribution of an embolus. *Med. Jahrb. d. Wiener Aerzte*, 1873, S. 195: 199.

Loring's important paper embodied all the arguments employed by opponents of embolism. The historic interest in Loring's paper consists in the etiologic factors, other than embolism, which he was the first to suggest as satisfactorily explaining the symptoms presented by his cases. Heart failure was ascribed as a cause for prodromal attacks of retinal ischemia which passed away when the heart recovered its tone and circulation was restored. In one case he suggested the presence of "some anatomically different condition which would make it more difficult (in this case) for the heart to force blood into the left eye than into the right; hence, recurring blindness always takes place in the left." Of another case he said, "May not the trouble have been caused by some morbid *local process* rather than by importation of a plug: Might not *thrombosis* occur in the retinal vessels and might not this attack be the first expression of a coming and more general trouble in the minute branches of the circulatory system at large?" The paper concludes as follows, "In the train of symptoms classed under the title of embolism, too much significance has been laid upon the stoppage of the circulation by importation of a plug formed at a distance from the eye, while not enough stress has been laid upon the mechanical actions regulating the supply of blood within the eye, as well as upon the *conditions of the walls of the vessels*."

There is nothing ambiguous in this language of Loring's in which he first systematically presented the arguments upon which he with Nettleship, Noyes and Priestly Smith founded the now dominant theory of a vascular origin for closure of the central artery.

will be able to force blood through the strictured lumen, and the phenomenon which has borne the name of embolism then occurs. After a time the blood pressure rises or the walls of the artery recede and circulation is restored. The damage to the retina will depend upon the length of time the blood supply was interrupted, which may have been minutes or days. This theory is very attractive and satisfactorily explains every feature presented by the great majority of cases. Endarteritis may cause complete or incomplete failure of the retinal circulation and, furthermore, permit the blood again to enter the retina. By accepting this disease as the cause of obstruction in the central artery we are enabled to understand the prodromal obscurations of vision, incomplete closure, sudden return of circulation and recovery from severe attacks. Successive embolic closures of one artery or coincident closure of both central arteries would require volleys of emboli, but there is nothing unreasonable in the assumption that a widely disseminated disease like arteriosclerosis may attack both central arteries or that periods of exacerbation may occur. In bilateral attacks recovery of one eye and continued blindness of the other are explained by the more advanced obliterative process in the blind eye. The supposed transfer of an embolus from a trunk artery to a branch, or from one point to another in the same artery, is explicable on the assumption that the ophthalmoscopic appearance of a migratory embolus was produced by several points of endarteritic obstruction at which the blood column was temporarily arrested.

In some cases microscopic examination has failed to reveal a plug in the central artery although the ophthalmoscopic picture indicated that this vessel was closed.\* The microscopic evidence of endarteritis as the cause of closure does not, however, depend upon failure to discover an embolus in the artery, but upon the interpretation of histological changes found in an actual obstruction.

UNUSUAL CAUSES. Béal reports a closure of the central artery following violent compression of the neck and thorax (*traumatic asphyxia*, p. 75). At first there was total blindness but eight days later the patient could count fingers. The fundus presented a typical picture of "embolism." The case terminated in optic atrophy.

Neuberger saw retinal ischemia follow a punctured wound of the orbit. Von Michel microscopically examined a traumatic case. The eye was injured by a cane. Profuse subconjunctival hemorrhage and exoph-

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\*Loring, Popp, Hirschberg, Michel and Uthoff.

thalamus followed. The fundus presented the characteristic picture of central artery closure. Six weeks later the patient died from tetanus. Near the optic foramen, a piece of wood was found embedded in fetid pus. A hyaline thrombus filled the lumen of the central artery from its origin at the ophthalmic to its bend after entering the nerve.

It has been suggested that the central artery frequently is closed by external pressure from various causes, such as tumors, swelling of retrobulbar neuritis, hemorrhage into the nerve or nerve-sheaths, etc. In most cases such pressure would interrupt circulation in the flaccid vein rather than in the rigid artery and the result would be stasis, not anemia. It is not infrequent, however, for the vein to leave the nerve shortly after passing out of the eyeball, the artery traversing the nerve alone for a distance of several mm. In such cases it is evident that pressure upon the nerve, behind the exit of the vein, would act only on the artery. Such a condition may have existed in Schweigger's case, in which a patient with unilateral, retrobulbar neuritis, suddenly became blind in the affected eye. The fundus presented the picture characteristic of "embolism." On the evening of the same day the arteries refilled and vision improved. Seggeŕ reported the case of a soldier whose left eye suddenly became blind from closure of the central artery. Death soon followed from pneumonia. The autopsy showed, on the distal end of the nerve, a point of firm adhesion between the nerve and its sheaths. Corresponding to this place, a small, firm tumor was found in the nerve. The growth was diagnosed as gliosarcoma.

In a few cases blindness with the ophthalmoscopic appearance of central artery occlusion has followed blunt force. In the cases of Nettleship and Oeller it resulted from a blow on the orbital bones. The probable cause was compression of the retinal artery near the ophthalmic by a fractured bone.

Closure of the central artery from hemorrhage into the nerve sheaths or into the substance of the nerve (Magnus) is unsupported by clinical or microscopic evidence.

**ANATOMICAL CHARACTERS.** In the optic nerve, the wall of the central retinal artery consists of three coats, *i. e.*, adventitia, media and intima. Of these, the intima is of the greatest pathologic importance. The *adventitia* is not separable from the connective tissue framework of the nerve. The *media* is composed of fibrous and elastic tissue containing a few muscle fibers. The intima consists of three layers, (1) a single layer of endothelial cells lining the lumen; (2) a layer of subendothelial

tissue; (3) a homogeneous elastic membrane, the *lamina elastica interna*, also called *crenated membrane* because it falls into folds when the artery is emptied.\* Nearly all the morbid changes in the central artery occur in the intima. As the retinal artery enters the eye it parts with its sub-endothelial tissue and elastic membrane. The latter, however, is replaced by elastic fibers.

Occlusion of the central artery usually occurs within, or near, the lamina cribrosa. Exceptionally, closure occurs in other parts of its course. In a case by Hirschberg the artery was closed just beyond its origin from the ophthalmic, outside the dural sheath. In Velhagen's case a plug was found at the point where the artery passed through the dural sheath. In the region of the lamina cribrosa the walls of the retinal vessels are more subject to disease than in any other part of the nerve or retina. The reason for this is obscure, although it is probable that the anatomy of this region is responsible. As the central vessels pass through the lamina they are somewhat narrowed† and firmly enclosed by condensed connective tissue. This constriction at the terminal end of the artery acts as a nozzle, giving added impetus to the blood stream just where it encounters resistance from intraocular pressure. A weak heart may be unable wholly to overcome this resistance and thus the blood may whirl and eddy at this point, a condition considered as sufficient to establish either a proliferative process or thrombus within the artery. Another factor is the abundance of perivascular tissue in this region, which, like connective tissue elsewhere, frequently becomes the seat of chronic inflammation. Haab has suggested that the movements of the eyeball, by dragging on the optic nerve, may provoke irritation where it is inserted into the globe.

An embolus excites inflammation in the walls of the artery both in front and behind its point of impact (Harms).

Toxic thrombi usually are of the red or hyaloid variety.‡ It is the

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\*The name, "elastic," which is applied to this tissue is unfortunate and leads to confusion, especially when considering the blood vessels whose elasticity and contractility are apt to be attributed to the action of *elastic* tissue. As truly stated by Coats, there is no proof that this tissue is elastic, in fact, it exhibits no elasticity as illustrated by the internal elastic membrane which becomes crenated on collapse of the artery. Furthermore, when the number of elastic fibres in the walls is increased the vessel loses all elasticity and becomes rigid.

†The normal central artery measures  $145\ \mu$  at the lamina and  $166\ \mu$  1 mm. behind it (Coats.)

‡When the blood is filled with pathogenic organisms, bacterial emboli may lodge in the central artery and produce occlusion. Früchte reports a case of sepsis with

bland, white thrombus which is supposed to close the central artery in those cases formerly considered as embolic. Unfortunately, the cases so far reported were not microscopically examined until the characteristic histologic structure of a thrombus had been lost in the process of organization. If an embolus or thrombus occludes the central artery, it is soon replaced by cells and tissue proliferated from the intima which are difficult to distinguish from similar cells and tissue produced by a primary endarteritis. This difficulty is increased after degenerative changes are established, at which time the original nature of the obstruction cannot be determined (Figs. 89-92). It is possible that calcareous or tissue masses resulting from endarteritic proliferation, may sometimes separate from the walls of the central artery and plug the vessel beyond the point of their origin.

A peculiarity of arteriosclerosis is its frequent, apparent limitation to a system of vessels or even a single vessel. As a rule the walls are affected for short distances only, uniform thickening throughout the course of an artery being rare. Nearly all the morbid changes in the central artery occur within the circle of the internal elastic membrane under the form of endarteritis obliterans or proliferans. The early histological changes occurring in closure of the central artery from endarteritis have not hitherto been reported. According to Ribbert and Jores, the most important preliminary process that takes place in arteriosclerosis is distension of the tissue and formation of spaces in the intima. Ribbert summarizes his views as follows: "As a result of increased blood pressure, fluid constituents from the blood are forced into the intima, separating the fibrillæ, dilating the spaces, and causing the fibrillated basement membrane to swell." In the following case the microscopic findings conform with Ribbert's description.\*

The patient was a man, 51 years of age. As demonstrated at autopsy, he had chronic endocarditis, pericarditis and pleurisy. The immediate cause of death was edema of the lungs. Two weeks before death the left eye suddenly became blind. The fundus presented the ophthalmo-

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ulcerative endocarditis in which closure of the central artery occurred. Longitudinal section of the nerve showed the lumen filled with a homogeneous, stratified mass, which Früchte considered an embolus, but others might regard as septic hyaline thrombus.

\*From the service of Prof. Francis Valk, N. Y. Post Graduate Hospital. I have withheld and studied this case for many years and the diagnosis was made only after the most careful research and consultation. Among others, Profs. T. Mitchell Pruden and Henry T. Brooks concur in the histologic findings described above.





Fig. 84.

Closure of Central Artery—Longitudinal Section.

The endothelial lining of the artery is compressed into a string of cells by serous exudate. In front of the occlusion thus formed, to the left in the figure, the lumen is filled with blood not possessing the anatomic structure of thrombus.

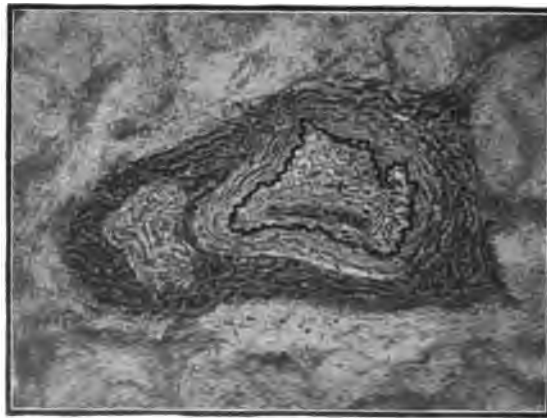


Fig. 85.

Closure of Central Artery by Active Endarteritis.  
The artery is shown to the right, the vein to the left.

scopic picture typical of an embolus in the central artery. The eye and optic nerve were removed at autopsy and sent to me for examination. The nerve was cut longitudinally. Complete serial sections were made and every section examined.

Immediately behind the lamina, the lumen of the central artery was closed by swelling of the intima. The swelling was due to a serous exudate in the spaces of the subendothelial tissue, which were so dilated that the endothelial lining of the artery was compressed into a string of cells (Fig. 84). The fluid which filled the subendothelial space was slightly fibrillated and contained a number of leucocytes but no red cells. Immediately in front of the occlusion the lumen of the artery was filled by old "stagnant" blood which did not possess the anatomic structure of a thrombus. Behind the occlusion the artery was almost completely collapsed and contained no blood corpuscles. The obstruction bore no resemblance to the endothelial detachments produced by rough manipulations (Fig. 90). Certainly, the exudate and leucocytes in the subendothelial tissue were not artefacts. In the later stages of endarteritis, two types of obstructive proliferation occur in the central artery: (1) cellular or cellulofibrous proliferation; (2) increase in the elastic tissue. These two processes frequently coexist. Mucous swelling, atheromatous softening, and ulceration do not occur in the central artery of the retina.

The following case is an exquisite example of the cellulofibrous type of endarteritis:\*

Man, 44 years of age, with advanced general arteriosclerosis. Entered the clinic with hemorrhagic retinitis. Glaucoma developed and the eye was enucleated. Two weeks later violent hemorrhage occurred from the stump.

The entire optic nerve was sectioned transversely and every section examined. The walls of the central vein were greatly thickened and completely collapsed. The lumen of the central artery was closed by active endarteritis (Figs. 85-86). In this case it is difficult to determine on which side of the vascular system occlusion first occurred. This difficulty is further increased by extreme angiosclerosis of the retinal vessels. Within the limits of the lamina elastica there is an exuberant proliferation of large vesicular cells (Fig. 86). Although epithelioid in type, these cells are not regarded as offsprings of the endothelium but of the subendothelial tissue (Coats). They either swell and disintegrate or become fibrous.

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\*From the clinic of Dr. J. Scott Wood, Brooklyn Eye and Ear Hospital.



The cut shows a mass of tissue formed in this manner, situated between the endothelial lining and nest of vesicular cells. The uniform appearance of the cells indicates that all are of nearly the same age. This fact suggests that an eruptive proliferation of such cells would close the artery, and later, through shrinkage, the lumen might reopen. In this as in all similar cases, proliferation proceeds from one side of the artery, producing an eccentric, irregular lumen. Frequently the cellulofibrous tissue assumes a nodular form which projects into the lumen usually from one side only, rarely from both (Fig. 81). In either case the lumen is reduced to a slit-like opening. At first the nodes consist of cells but later the mass is invaded by elastic and fibrous connective tissue, which replace the cellular elements (Fig. 88).

More or less increase of elastic tissue in the intima occurs in all types of endarteritis, but sometimes proliferation of this tissue is so excessive as to constitute the sole, or at least principal, cause of arterial obstruction. The vessels shown in Fig. 87 are from a case of general arteriosclerosis. The section was made through the anterior part of the lamina cribrosa and stained by Weigert's method for elastic tissue. Both branches of the central vein are completely closed by cellular and fibrous tissue. The intima and media of the central artery are replaced by thick layers of elastic tissue. As a rule, the media takes no part in the sclerosing process. Occasionally, however, as in this case, concentric narrowing of the lumen occurs from proliferation of elastic tissue in the outer coats which replaces the muscularis as well as the intima. In the illustration, notice the entire absence of elastic fibers in the veins, which proves that these vessels have already assumed their retinal structure. In the following case the central artery was entirely replaced by a cord of elastic tissue.

Male, 32 years of age. Operation on the frontal sinus was followed by orbital cellulitis. I saw him first at this time, in consultation with Dr. Lewis Coffin. O. D. Exophthalmus was excessive. Cornea hazy. No perception of light. Blindness occurred suddenly. Details of fundus were obscured by condition of the cornea. Pupil dilated and immobile. Vitreous clear. The fundus had a uniform, dirty-white reflex. No retinal hemorrhages. View of the vessels was unsatisfactory. An opinion was given that the central artery was closed by septic thrombosis. Incision of the orbital tissue evacuated pus and the exophthalmus gradually subsided. The eye never recovered perception of light. Ulceration and cicatrization of the cornea prevented further examination of the fundus. Three months later the eye was enucleated. Transverse serial sections were

**Fig. 86—Closure of Central Artery by Endarteritis. The Artery is to the left in the Figure. Within the Elastic Lamina, which is Crenulated and Deeply Stained, a Proliferation of Large, Vesicular Cells has taken place.**

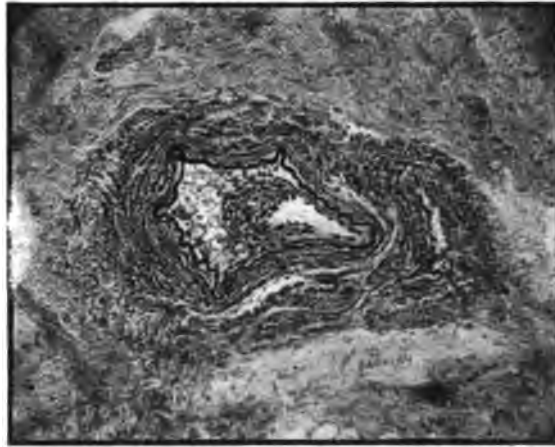


**Fig. 86.**

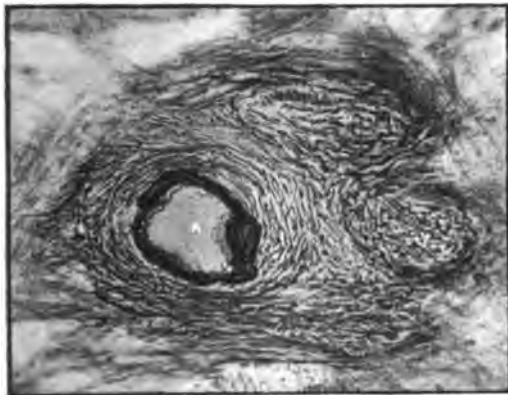


**Fig 87—Complete Closure of both Branches of the Vein (to the right) by Fibrous Tissue. The Artery (to the left) is Narrowed by Proliferation of the Media, its Elastic Lamina is Deeply Stained by Weigert's Method.**

**Fig. 87.**

**Fig. 88—The Obstruction caused by Endarteritis has been Converted into Fibrous Connective Tissue in the Vein (to the left) and into Elastic Tissue, which is Deeply Stained, in the Artery, shown to the right. The Vein is hardly Distinguishable from the Surrounding Connective Tissue.**

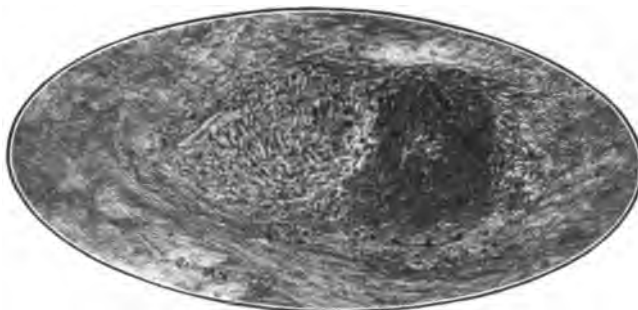


**Fig. 88.**







**Fig. 89.**

**Occlusion of the Central Artery.**

**Degenerative changes render impossible the determination of the true nature of the obstruction.**

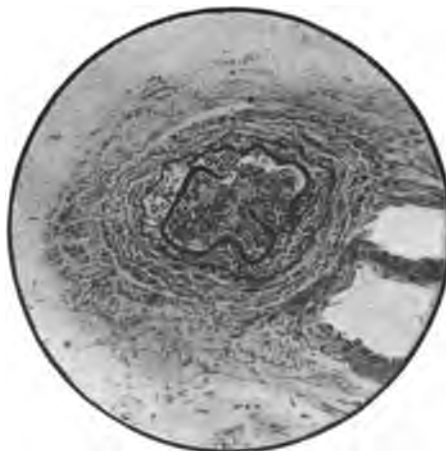


**Fig. 90.**

**Occlusion of the Central Artery.**

**The Endothelial Detachment is an artefact produced in the preparation of the specimen. The elastica (deeply stained) has been stripped from the walls.**

made of the posterior part of globe and optic nerve. Both central artery and vein were completely obliterated in and behind the lamina cribrosa. The two vessels were enclosed in a common sheath which was moderately infiltrated with round cells. The artery consisted almost wholly of elastic tissue (Fig. 88) and the vein of organizing connective tissue and cells. The retina was edematous and its inner layers degenerated. No evidence of intraocular hemorrhage was found except some pigment granules on the disc.

A frequent artefact in the central artery, often mistaken for a pathological process, is produced during enucleation by dragging and torsion of the nerve and especially, by unsuccessful attempts to sever the nerve with scissors. A great variety of conditions result, but in all, the distinguishing feature is a fresh, normal condition of the blood and tissue elements. Usually the lumen of the artery is blocked by massed blood corpuscles and spindle cells. The endothelial layer may be shredded or the elastica stripped from the walls and entangled in the blood (Fig. 90). Collections of fresh blood may occupy the intima. Cases reported as detachment of the intima, exfoliation of endothelium, dissection of the walls by blood and fresh thrombi are examples of this artefact. The only case of active endothelial proliferation and loosening which I have seen was also due to traumatism, but in this case it is believed to have occurred before enucleation.

L. M., healthy mechanic 27 years of age. The left eye was perforated by a piece of steel which passed through the ciliary body and lodged in the tissues at the inner border of the papilla. Intraocular hemorrhage was slight. Four hours later the steel was located by skiography and removed by magnet. Mild cyclitis developed and, fearing sympathetic ophthalmitis, the eye was enucleated on the fourth day after injury. Microscopic examination showed no evidence of purulent infection. The former site of the foreign body was distant about 1 mm. from the central artery. There was round cell infiltration of the surrounding parts but the tissue process appeared to be reparative rather than inflammatory. The following interesting changes were noted in the central artery. Distension of the perivascular lymph sheath and, on the side of the vessel adjacent to the injury, there was edematous thickening of the media. At this point the endothelial cells were loosened and increased in numbers, forming a loosely adherent mass which projected across the lumen. Adjoining this mass, were single cells, partly detached and adherent to the walls by pointed tips, their long diameters extending out into the lumen (Fig. 91).

In endarteritis, the progressive deposit of new tissue finally is arrested by nutritive disturbance and retrogressive alterations ensue. If, through these processes, the central vessels are occluded, the products of proliferation and degeneration may be removed and all traces of the vessel obliterated by encroachment of the surrounding connective tissue (Fig. 92).

CLINICAL HISTORY. Prodomata, consisting of transitory obscurations of vision, which may be alarming or so slight as to be forgotten by the patient, usually are present. At last, sudden blindness ensues which is permanent. Transitory attacks may continue for twenty years or longer with no apparent injury to vision. In other cases, permanent blindness may follow the first seizure. An attack usually is unattended by subjective symptoms, although vertigo, faintness and photopsia have been observed. In rare instances both eyes are affected simultaneously or at different times.\* In Hasse's case the interval was ten years. In bilateral cases the eyes may be affected in varying degrees. After a severe attack most cases advance to complete blindness, even though temporary improvement may take place. In a case observed by Sichel there was at first great improvement, but four months later vision again sank to bare perception of light. If the blindness is absolute during an attack, the prognosis is bad. If central scotoma exists, useful vision cannot be expected. At first the whole field may be darkened and later a quadrant clear up. Even in the most favorable cases peripheral contraction of the visual field is to be expected. A prognosis of useful vision may be given in cases where central vision is preserved. Despite the unfavorable outlook for most cases, a number have been reported in which vision was nearly or completely restored.†

A few cases have occurred and been microscopically examined, in which closure of the artery was followed by unilateral glaucoma. In all but three of these the increased tension was explained by pathological conditions other than closure of the artery.‡ Closure of the central artery is not very rare, and three cases occurring in connection with glaucoma constitute too small a percentage to establish a causative connection between the two conditions. It is held that arteriosclerosis is the common

\* Hirschberg, Ewers, Hasse, Benson, Blagowjestschenski.

† Uthoff, Raehlmann, Mooren, Olaf Page, Coats.

‡ Cases of Nettleship, Manz and Marple. Nettleship's second case is not included, owing to lack of correspondence between the clinical and microscopic findings. The onset of blindness was attended by severe pain and the ophthalmoscopic picture was not characteristic of "embolism." Probably, the pathological examination was not complete.







Fig. 91.

Fig. 91—Edothelial Proliferation due to Traumatism. The Perivascular Lymph space is Distended, the Media Thickened, and the Endothelial Cells are Increased in Numbers and Project across the Lumen.



Fig. 92

Fig. 92—Degenerative Changes following Obstruction and Obscuring the Nature of its Origin. All Traces of the Vessel have been Obliterated by Surrounding Connective Tissue.

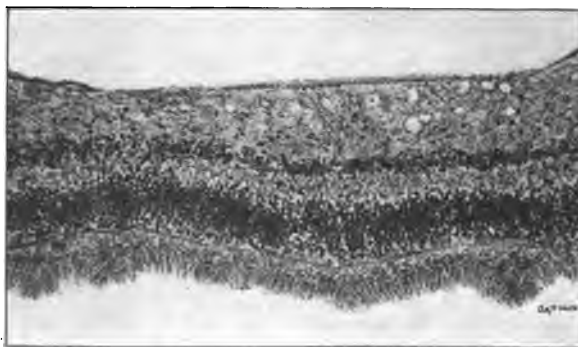


Fig. 93.

Fig. 93—Retina Two Weeks after Closure of Central Artery. The Inner Layers are Degenerated and Edematous.

cause of both the central artery closure and subsequent glaucoma. Were this true, the order of events occasionally would be reversed and closure of the artery follow glaucoma; such an occurrence, however, appears to be unknown.\* Primary glaucoma is regarded as a bilateral affection. In these cases the glaucoma was limited to the eye in which the artery was closed, indicating that it was secondary to some morbid condition in the affected eye. That cessation of retinal circulation would directly affect intra-ocular pressure, is inconceivable; therefore, the development of glaucoma, probably depended upon some local process which was not ascertained. It may be possible that, inasmuch as the inner layers of the retina die after closure of the central artery, the products of their decomposition may excite sufficient reaction in the pectinate ligament to establish a glaucomatous tendency. It has not been established, however, that such a tendency exists.

**DIAGNOSIS.** Recent, well developed cases of central artery closure can hardly be mistaken for any other condition. If the retinal opacity is very great, it may completely obscure the nerve. Such cases are differentiated from choked disc by the small veins and absence of swelling; also by the macular spot and by amaurosis. The foveal red spot is nearly diagnostic. It may develop also in *comotio retinæ*, but in this condition the vessels are not reduced in size. Amaurotic idiots show a similar appearance at the macula, but there is no opacity around the nerve and the retinal vessels are normal. Amaurotic idiocy occurs only in infants. A macular hole sometimes appears in the retina after injury, especially in myopic eyes. In these cases peripheral vision is preserved, while in closure the entire field is darkened. Small macular hemorrhages are usually round and may resemble the red spot of "embolism." After circulation is resumed and vision partly restored, the diagnosis rests largely on the history. At this time, slight pressure on the globe with the finger may arrest retinal circulation and render visible obscure mural opacities. In old cases of closure in which circulation has been resumed, concentric contraction of the visual fields usually exists.

**OCCULSION ATROPHY (Atrophic Stage of Central Artery Closure).** If the central artery is once completely blocked by *endarteritis obliterans*, embolus or thrombus, the inner layers of the retina and the optic nerve undergo extreme atrophy (Figs. 93-94).

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\* Ridley's case, which Haab quotes as an example, was one of hemorrhagic glaucoma, in which wide spread vascular disease existed.

**STEREOGRAM 44. RETINAL ATROPHY FOLLOWING CLOSURE OF THE CENTRAL ARTERY.** Left fundus oculi of girl who two years ago had complete closure of the central artery of the retina. The ophthalmoscopic picture which the eye presented at that time is shown in stereogram 41. Vision never was restored after closure first occurred. Externally, the eye appears normal. Light perception is absolutely abolished. Direct pupillary reaction to light is lost but consensual reaction is normal. The fundus picture is characteristic of atrophy confined to the inner layers of the retina. There is no pigmentation or other evidence that either the outer layers of the retina or the choroid are in any way disturbed. The atrophic nerve has a bluish cast but the markings of the lamina cribrosa are not visible, as is the case in simple optic atrophy. Only two arteries contain blood columns, the others being reduced to white bloodless cords or entirely obliterated. The veins which remain are highly sclerosed, but contain more blood than the arteries. A delicate veil, representing the atrophic layers of the retina, hangs over the entire fundus.

**DIAGNOSIS.** Occlusion retinal atrophy is recognized by its extreme character, absence of postinflammatory products, pigmentation or hemorrhage and by absolute blindness.

**BRANCH ARTERY CLOSURE.** When a branch of the central artery of the retina is obstructed, the same changes occur within the area of its distribution as in obstruction of the trunk artery and a scotoma, corresponding to the affected region, appears in the visual field. It has been stated that the macular branch is most frequently closed. Probably, this is more apparent than real, inasmuch as a small, peripherally situated scotoma may escape detection, whereas, a central defect is noticed at once. Closure of a sub-branch causes a fan-shaped scotoma highly characteristic of the condition. The retinal opacity is more prominent in branch closure than in closure of the central artery, from contrast with the adjacent, unaffected retina. Occasionally, edema is excessive. Hirschberg, describing such a case, said: "Throughout the area of distribution of the obstructed artery the retina was whitish and edematous. To those unacquainted with the picture it might be mistaken for detachment of the retina, a mistake that appears sometimes to have occurred." It must be remembered that the retinal opacity soon passes away, after which the visible alterations may not correspond with the functional disturbance. At this period perimetric measurements will reveal the characteristic scotoma of branch closure, thus indicating the real trouble. By the same means it may be possible to distinguish a primary vascular degeneration,

from a vascular degeneration secondary to closure of an artery. Bleeding usually is absent in closure of a branch artery. An associated branch artery and vein, however, may both be closed from endovasculitis or thrombosis, in which case hemorrhages and edema occur throughout the region of their distribution. The possibility of hemorrhagic infarction from incomplete closure of a branch retinal artery is a subject for discussion (see p. 112). Reimer considers branch closure usually to be incomplete because a broken blood column has never been observed on the peripheral side of the obstruction.

The point of obstruction being ophthalmoscopically visible in branch closure, the latter has been subjected to the most careful scrutiny. Many observers have seen what they believed to be an embolus blocking the artery. These obstructions have been variously described as "dark-red enlargements," "white or grey lines," or as "white scales on the vessels," etc. Sometimes the plug has appeared to change its position, spontaneously or from external pressure. Frequently, they have disappeared altogether. The more complete knowledge acquired regarding the ophthalmoscopic appearances produced by arteriosclerosis makes it more than probable that nearly all so-called emboli, seen with the ophthalmoscope, in reality, were illusory effects incident to changes in the walls of the arteries. Thus, sclerotic spots in an otherwise transparent vessel, or transparent places in a sclerosed vessel, or a point of obstruction at which the blood column was arrested, were misinterpreted if the mind of the observer was possessed by the embolic theory. A conical pointing of the blood column at each end of the supposed embolus has often been described, a condition now known to be pathognomonic of narrowing of the lumen from thickening of the walls. Raehlmann, speaking of the ophthalmoscopic appearance in arteriosclerosis, says: "In former years, when I first observed these changes, I thought I had before me the remains of an embolic process." From the evidence at hand it must be concluded that so-called branch embolism is, in the great majority of cases, due to obstruction of the blood stream from thickening of the arterial walls.

STEREOGRAM 45. CLOSURE OF BRANCH RETINAL ARTERY. Married woman, 30 years of age. Mother of two healthy children. Has never miscarried. Wassermann test for syphilis and Von Pirquet for tuberculosis, negative. Urine contains a trace of albumin. Blood pressure 125 mm. Hg. Dr. Glentworth Butler, who made a physical examination, found neither cardiac nor vascular disease. Four days ago sudden blurring occurred in the left eye. Light perception is lost in the upper-inner visual field (Fig.

95). With the ophthalmoscope a retinal opacity is seen covering the infratemporal quadrant of the fundus, most dense in the macular region. On the temporal side, the opaque and normal retina are sharply separated by a wavy line which passes directly through the fovea centralis. On the infranasal side, the white opacity fades gradually into the normal red fundus. The opaque region is triangular in form with the apex at the disc. The lower half of the fovea is situated within the opacity where it appears as a semicircular red spot, continuous above with the normal macula. The affected region represents the area of distribution of the infratemporal retinal artery which is obstructed at the disc. At its exit from the nerve this vessel is opaque for a distance of about one disc diameter. Within the opaque portion of the artery are two, short, red segments of visible blood column. Branches of the artery distributed to the macula are rendered visible by contrast with the opaque retina. The infratemporal vein contains blood but is broader at the periphery than at the disc. Evidently, the arterial occlusion is incomplete. Pressure with the finger on the eye fails either to empty the occluded artery of blood or produce pulsation.

**DIAGNOSIS.** Closure of a branch artery often escapes discovery. It may be mistaken for flat detachment of the retina or edema due to effusion from the choroid. In detachment both arteries and veins are full, darkened and somewhat tortuous, while in arterial occlusion the vessels are straight and narrowed. The surface of a detachment always is somewhat elevated, which is not the case in occlusion. Good light perception or even vision is retained in recent detachment, but the area affected by closure of an artery is blind. Moreover, the triangular form of opacity in branch closure is diagnostic of the condition. An isolated patch of opaque retina from choroidal effusion (Stergm. 18), is differentiated from the opacity of vessel closure by its elevated surface, normal or congested vessels, its form and situation, which bear no relation to an artery and, in nearly all cases, the presence of other choroidal lesions. Photopsiæ are a symptom of choroiditis but are not present in closure of the retinal vessels. Circulation may be restored after branch closure, but the scotoma may remain. The triangular shape of such scotomata indicates their vascular origin.

#### *Bibliography.*

- Alexander, Bericht u. d. 25. Versam., d. Oph. Gesel. z. Heid., 258.  
 Béal, Annal. d'Ocul., Aug. 1909.  
 Behse, A. f. O., LXX, 2.  
 Benson, O. H. Rep., X, 336. Trans. Intern. Oph. Cong., Edinbg., 1894, p. 81.  
 Bernheim, Jour. Am. Med. Assn., LV, 4, July 23, 1910, 283.

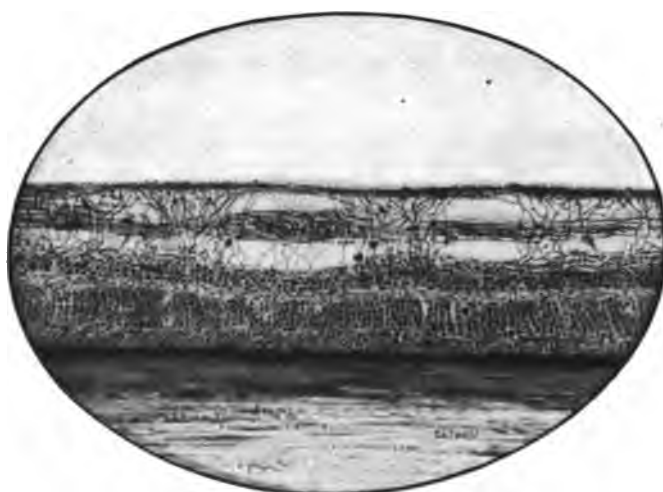


Fig. 94.

Retina after Closure of the Central Artery.

The inner layers are markedly atrophic.

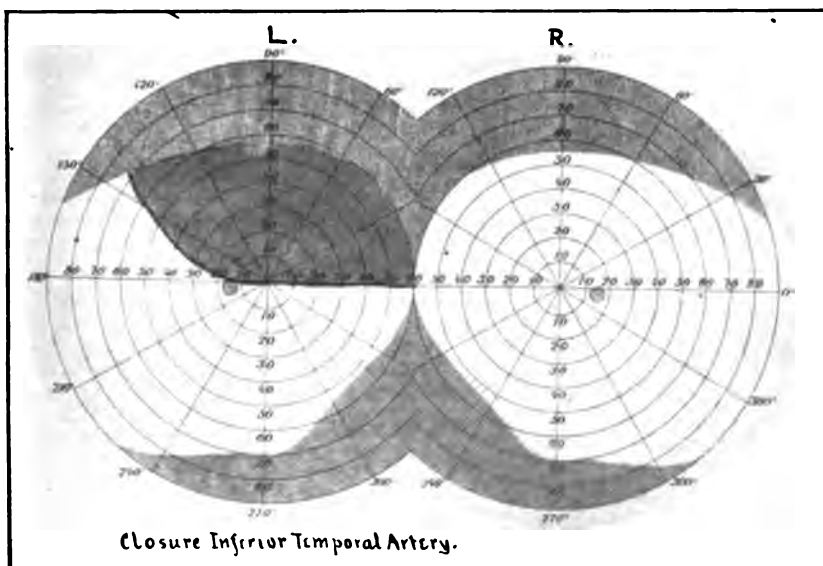


Fig. 95.



- Blessig, A. f. O. VIII, 1866, 320.  
 Bull, *Öle Krankheiten der Retinalgefäße*. Leipzig, 1903.  
 Blagowjestschenski, ref. *Jahrsber. f. Oph.*, 1872, 342.  
 Coats, O. H. Rep., XVI, 1, 1904; XVI, 4, 1906. T. O. Soc., XXIV, 1904, 163.  
 Dogiel, Arch. f. mik. Anat., Taf. II, Bd. XL, 1892.  
 De Schweinitz, Rep. Phil. Path. Soc., 1897-99. Ophthalmology, 1906, 652; Arch. of Oph. Mar. 1910, 101.  
 De Bono, Arch. di Ottalm. 2, 1894, 238.  
 Dotto, Arch. di Ottalm. 1, 1894, 234.  
 Elschmig, A. f. O. XLIV, 1, 1897. 144 Kl. M. f. A. 1898, 93; A. f. A. 1892, 93.  
 Ewers, *Jahrsberch. f. Oph.*, 1899, 661 (Ref.).  
 Faravelli, Annl. di Ottalm. XVI, 1887, 63.  
 Fischer, Über Emb. d. Art. Cent. r. Leip. 1891. Deutsch. m. Woch. Früchte, Kl. m. f. Augk. 5, 1908, 245.  
 Fuchs, Text Book.  
 Gonin, A. d'O., XXV, 1905.  
 Grüning, A. f. A. VI, 1877, 35.  
 Gunn, Trans. Oph. Soc. United Kingdom, 1895.  
 Haab, Norris & O. IV, 481. Korespbltz. f. Schwitz. Artze, 1898, 2.  
 Harms, A. f. O., LXI, 1905, Heft 1, p. 1; Heft 2, p. 245. Coresponbltz. 6 Schwitz. Artze, 1899, 338.  
 Hasse, A. f. A., X, 469.  
 Herter, C. f. p. A., 1879, 229.  
 Hirschberg, Centb. f. p. A., VIII, 70; IX, p. 43; 1882, p. 327; 1885, p. 38; 1885, p. 353; 1888, p. 296; 1889, p. 202; 1890, p. 322; Berlin, Kl. Wochft., XXII, 1875.  
 Hurd and Holden, N. Y. Med. Rec., July 11, 1903.  
 Inoyu, Centb. f. p. A., 1892, 320.  
 Jeger, Starr u. Starroperation, 1854, 194-8. Nagles Jahrb., 1876, 55.  
 Jores, Wesen u. Endwick. d. Art. Wiesbaden, 1903.  
 Kern, Inaug. Dissert. Zurich, 1892.  
 Knapp, A. f. A., X, 1880; T. O. Soc., XIV, 1904, p. 172.  
 Knies, Bericht d. Oph. Gesell. Heid., 1897, p. 162.  
 Leber, Bericht d. Oph. Gesell., Heidelberg, 1897, 162.  
 Logetschnikow, Ref. in Jahresber. f. Oph., 1904, 636.  
 Loring, Am. Jour. Med. Sci., 1874, 313.  
 Magnus, Die Schnervenblutungen, Leipzig, 1874.  
 Manz, Festsft. f. von Helmholtz, 1891, p. 9.  
 Marple, N. Y. Infirm. Rep. 3, p. 1.  
 Mauthner, Wien Med. Jahrb., II, 1873, 195; Lehrbuch der Ophthalmoskopie, Wien, 1868; Ref. in Nagles Jahrb., 1873, 337.  
 Merkle, Handbuch, Bd. I, 1887.  
 Michel, Jahrsber. f. Oph. 1898, 299-302; A. f. O., XXIV, 2, 1873; Z. f. A. XXI, 1909, 116; Z. f. A., II, p. 8.  
 Mintz, Zentrbl. f. Chirg., 1905.  
 Mooren, Ophthal. Mitteil, 1873, p. 89.  
 Nettleship, Br. Med. Jour., 1879, I, p. 889; Oph. H. Rep., VIII, 1874, p. 9; Oph. H. Rep., VIII, 1875, p. 251; Oph. H. Rep. XI, 1887, p. 268; Helmholtz Festsft., 1891; Michel's Jahrbbericht, 1895, p. 528; Nettleship & Lawford, T. O. S., 1882.  
 Neuberger, Munich Med. Woch., 1901, 1550.  
 Noyes, T. Am. Oph. Soc., 1888, p. 98.  
 Oeller, Atlas Selt. Oph. Erkrankung, C. Tab. XIV  
 Oglesby, Ref. in Wilbrand u. Saenger, IV, 1, p. 352.  
 Page, Olaf Am. J. Med. Sci., LXVII, 1874, 126.  
 Panas, maladies des yeux, 1894.  
 Parisotti, Annal de Ocul., CXIX, 321.  
 Parsons, Path. of The Eye, Quinine, IV, 1337; Oph. cil. neurec., 1187.  
 Pearse, Albany Med. Annals, June, 1901, p. 309.  
 Pearls, Centb. f. p. A., 1892, 161. Piersol, Nor. & Oliver, I, 327.  
 Popp, Inaug. Dissert., Wurtzburg, 1875.



- Quaglino, *Annal. d'Ocul.* LXV, 1871, 129.  
 Raehlmann, *Zeits. f. Klin. Med.* 1889, 606; *Fortsch. d. Med.*, 1889, 928.  
 Ramorino, *Annali di Ottal.*, VI, 25.  
 Raynaud, *Arch. gen de Méd. Janv. et Fév.*, 1874.  
 Recklinghausen, *Handb., allg. path.*, Stuttg., 1883.  
 Reimer, *Versam. d. Heid. Oph. Gesellsch.* 1898; *A. f. A.*, XXXVIII, 1898, 209, also p. 291; *Arch. of Oph.*, XXXII, 1903, 552.  
 Ribbert, *Ver. d. Deutschen Path. Gesellsch.*, 1904, Heft 2, p. 168.  
 Ridley, *Oph. H. Rep.* XIV, 1895, p. 264.  
 Rohmer, *Anls. d'Ocul.*, 134, p. 163.  
 Rothmund u. Eversbusch, *Mitteil.*, a. d. Universitätsaugenklinik zu Munich, 1, 328.  
 Schapring, *C. f. p. A. Dec.* 1906. Schlodtmann, *Über d. Extirp. Retrob. Tumor. m. Erhal. d. Aug. etc.*, Case 1 = alle, 1900.  
 Schmidt, *A. f. O.* 1875.  
 Schmidt—Rimpler, *A. f. O. Bd.* XXI, 1875.  
 Schnable und Sachs, *A. f. A.* XV, 1885.  
 Schnable, *A. f. A.*, XV, 1885 337; *A. f. O.* VIII, 1, 1862; XXVI, 1, 1880, p. 1.  
 Schnellcr, *A. f. Oph.*, VIII, 1, 1862, p. 271; *A. f. O.*, 1, 1880, p. 1.  
 Schweigger, *Vor. d. Gebrauch d. Augenspieg.*, 1864, p. 138, pl. 3, fig. 10; *Handb. d. Spec. Augenheilk.*, 425-515; *A. f. A. Bd.* XI, p. 144; *Arch. of Oph. N. Y.*, XXX, 1901, p. 506.  
 Seggle, *Bayr-ärztl. Intelligen.*, 1870, 13-14.  
 Shiba, *A. f. Oph.* LXIII, 373.  
 Sichel, *Annal d'Ocul.* LXVII, 314.  
 Siegrist, *Mitteil. aus Klin. d. Schweiz.*, 1, Heft X, 1894; *A. f. O. L.*, 548.  
 Silberstein, *Munich Med. Woch.* 1906, 626.  
 Smith, *Priestly Oph. Rev.* III, 1884, 463; *Brit. Med. J. Apl.* 1874, 452.  
 Steffan, *A. f. O.* XII, 1866, p. 34.  
 Strong, *The Laryngoscope.* St. Louis, Feb., 1911.  
 Uthoff, *Berlin Klin. Woch.* 1890, 825 and 1906, 626; 27 *Berich. d. Heid. Oph. Gesell.* p. 121.  
 Valaude, *Annal. d'Ocul.*, CXII, 266.  
 Velhagen, *ref. in Jahresb. f. Oph.* 1905, 240.  
 Von Graefe, *A. f. Oph.*, V., 1859, p. 136.  
 Von Michel, *Zeit. f. A.*, XXI, 1909, 116.  
 Waggenmann, *A. f. O.* XLIV, 2, 1897, 219 and XLIV, 3; XXXVI, 4, 1890, p. 1; XXXVIII, 3, 1892, p. 213; XL, 3, 1894, p. 221.  
 Weil, *Presse Med.*, Oct. 1905.  
 Welch, *Trans Assn. Am. Physicians*, 1887; and in Albutt and Rolleston, VI, 1909, p. 762.  
 Welt, *A. f. A.*, XLI, 335; *A. of O. N. Y.*, XXX, 1901, 495.  
 Wilbrand u. Saenger, *IV. Path. d. Netz.* 1909, p. 273, also vol. III, p. 664-786.  
 Zarhelli, *Beitrag. z. path. Anat. u. z. alleg. Path.*, XLVII, 3, 1910, p. 539.

#### CLOSURE OF THE CENTRAL VEIN OF THE RETINA (Thrombosis).

When the central vein of the retina is occluded, the conditions existing in closure of the central artery are reversed. Instead of retinal ischemia, a state of static, venous engorgement ensues, the intensity of which depends upon the degree of obstruction in the vein. Michel, who first demonstrated the condition, separates it into three classes: (1) Complete obstruction; (2) Nearly complete obstruction; (3) Partial or slight obstruction.

In cases of the first class, namely, complete obstruction, the fundus presents the picture of extreme hemorrhagic retinitis. The veins are

enormously distended, irregular in size and very tortuous, the sinuosities being mostly perpendicular to the plane of the retina, rising above the surface in black, knuckle-like loops and then disappearing into the swollen retina or pools of blood. The arteries are more or less diminished in size, according to the amount of pressure exerted upon their walls by swelling of the nerve.\* The arteries about the disc frequently are obscured by blood and exudates. The vessels, especially the veins, may be bordered by white lines due to wall degeneration although tortuosity and bleeding are not great when the veins are highly sclerosed. White borders to the veins may be due also to effusion into the perivascular lymph channels (perivasculitis).

The retina is covered with flame-shaped or round hemorrhages, most numerous around the disc, where the blood may form a continuous sheet. As a rule, the blood does not break through the limiting membrane of the retina into the vitreous. Areas of white exudation abound, especially where sharp bends occur in the veins. The macula exhibits a grey discoloration unless, as is frequent, it is the site of hemorrhage. There is always more or less redness and edema of the nerve-head. Usually the swelling is very slight, but occasionally is great enough to be classed as neuritis (*hemorrhagic neuroretinitis*). I have usually found the nerve swollen when retinal thrombosis was associated with albuminuria. Redness of the nerve likewise is variable, sometimes slight and again sufficient to obliterate the outline of the disc. If the central vein is occluded, the veins upon the disc cannot be emptied by external pressure upon the globe, although an intermittent blood current can be thus produced in the arteries, indicating that, notwithstanding closure of the trunk vein, circulation through the venous collaterals in the nerve still exists. In a case of Balaban's, the veins could not be emptied by pressure, but circulation was manifested by the unusual feature of a granular current in the veins.

In cases of the second class, in which *nearly* complete obstruction exists, the veins are equally dilated but hemorrhages are less numerous than in the first class. Probably, the veins upon the disc can be emptied by external pressure.

In cases of the third class, where obstruction is comparatively slight, the veins still are quite tortuous and the disproportion in size between them and the arteries is very apparent. Hemorrhage is slight or may not occur. The veins on the disc can be seen to empty when pressure is applied to the eye.

\*Reimer attributes the shrinking of the arteries to obstructive endarteritis.

Wilbrand and Saenger state that bleeding in venous obstruction may be slight under the following conditions: (*a*) extreme weakness of the heart; (*b*) incomplete closure of the vein; (*c*) closure of both central artery and vein; (*d*) when cilioretinal veins are present.\*

In the early stage of venous closure the retinal exudates have a milky, edematous appearance, due to large admixture of serous transudate. The dense white patches which appear a few days later are connected with the processes of blood resorption.

STEREOGRAM 46. CLOSURE OF CENTRAL RETINAL VEIN. (Retinal Thrombosis; Retinitis Hemorrhagica; Apoplexy of the Retina). Mulatto woman, 45 years of age. Laundress. Urine normal. Blood pressure, 250 mm. Hg. Surface arteries, rigid. Numbness of left hand and leg. Five weeks ago, on waking in the morning, she discovered that her left eye was blind. At the present time, with this eye, she can see movements of the hand. The ophthalmoscope shows the entire fundus covered with blood. Most of the hemorrhages are striate but many are deep and round. An almost continuous sheet of blood extends around the upper and temporal sides of the disc. The nerve head is not swollen, but is covered and obscured by a patch of white exudate. Similar masses of exudate are scattered throughout the fundus. The enormously dilated and tortuous veins are either entirely buried in the swollen retina or rise above the surface in dark, venous loops. The arteries are also obscured by the swelling and hemorrhage, but wherever visible they do not appear narrowed. The veins cannot be emptied of blood by external pressure on the eye.

DIAGNOSIS. Only the condition known as thrombosis of the central vein of the retina produces the picture shown by this stereogram. From choked disc it is differentiated by the extensive bleeding and absence of nerve swelling. Hemorrhage from traumatism would be more local or, if extensive, blood would appear in the vitreous. That venous obstruction in the above case is complete is shown by the amount of blood extravasated and by inability to empty the veins on the disc by external pressure with the finger.

ETIOLOGY. Aside from albuminuric cases, closure of the central vein usually occurs in elderly people who, although apparently in good health, in reality are subjects of cardiac or vascular disease. The disorder is prone to attack those who have passed their lives in laborious toil, espe-

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\*The existence of cilioretinal veins is doubtful (page 16).

cially in a stooping position.\* The disease is more common among men than women which may be attributed to the more violent exercises or intemperate habits of the former, conditions which lead to degenerative changes in the heart and blood vessels. The most frequent systemic change noted has been arteriosclerosis, rigidity of the surface arteries being recorded in most reports. Old age predisposes to the affection. Thus, among 54 cases tabulated by Harms, 44 were over forty years of age.† The average ratio of predisposing and associated diseases is illustrated in 50 cases collected by Moses of complete obstruction in the central vein or some of its branches. Among these, 38 had arteriosclerosis. The majority were over 50 years of age. In those below this age, other morbid conditions coexisted with vascular disease. For example, 5 were diabetic, 4 had contracted kidneys and 2 pernicious anemia. Two cases showed struma and mitral insufficiency. In one case the heart was displaced by kyphoscoliosis. The influence of vascular disease upon the retinal circulation was illustrated in a case where the central vein was closed in one eye and the central artery in the other. In another case of closure the fellow eye exhibited the picture of albuminuric retinitis.‡ In closure of the central vein, disease of the retinal vessels is usually found on microscopic examination, although the degenerative changes may not have been sufficiently advanced to produce visible opacity of the walls when viewed with the ophthalmoscope. In Schönewald's 18 cases, retinal angiosclerosis was ophthalmoscopically observed in nine. Syphilis is rarely mentioned as a causative factor, but should be carefully sought for when retinal thrombosis appears in young people. Nephritis and diabetes, by producing vascular degeneration, are causes of thrombosis but when, as is common in these diseases, thrombosis occurs in a branch vein, it is associated with such wide-spread inflammatory and exudative conditions that the ophthalmoscopic picture is classed as retinitis of albuminuric or diabetic origin. Thrombosis is not uncommon after influenza and in a considerable proportion of cases it forms in the central vein.§ In other microbic and exhausting diseases thrombosis of the central vein is rare, (marantic thrombus). Würdemann saw a case following mumps, in a

\*Among Amman's 20 cases, 13 were accustomed to gardening and 2 were smiths. Haab also mentions posture as exerting an influence upon thrombus formation.

†Goh's case of septic thrombosis is omitted.

‡Schönewald collected 18 cases of thrombosis of the central vein. There was general arteriosclerosis in 7, grave heart disease in 3 and pulmonary emphysema in 2. Four developed glaucoma and one died from cerebral apoplexia.

§Three of Coat's 16 cases followed grip. In cases of intraocular trouble following grip, the accessory nasal sinuses should be carefully examined.

boy eight years of age. Goh examined the eyes of a man 25 years of age who died from "sepsis hemorrhagica." There was a hyaline thrombus in the right central vein. In the left eye, several retinal vessels and a choroidal vein were closed by thrombi.

Thrombosis is favored by certain changes in the composition of the blood, particularly anemia. In pernicious anemia, cases are reported by Moses and Michel. Both Bondi and Michel have seen cases in leukemia. The one serious complication of chlorosis is thrombosis and sometimes this occurs in the central vein (Clermont, Ballaban). Haab has observed retinal thrombosis in the intense anemia of pregnancy. Michel thinks that incomplete thrombosis of the retinal vein is common in the cachectic anemia of malignant growths.

In orbital cellulitis closure of one or both central vessels occurs from thrombosis or proliferating vasculitis. These cases are attended by intense inflammation and exophthalmus. They are of microbic origin and originate in accessory sinus disease, facial erysipelas, wounds of the orbit, etc.

An abnormal course of the central vein predisposes to sclerosis and thrombus formation. In 25% of the cases examined by Harms the central vein formed loops before emerging from the nerve sheath. Tortuosity of the vein also is described by Goh, Welt, Yamaguci and Coats.

Other conditions which have been supposed to cause thrombosis of the central vein are: suppression of menses; menopause; cessation of customary hemorrhoidal bleeding; extensive burns of the skin; poisoning by phosphorous and by nitro-benzol. In the condition last named the fundus presents a violet color and the blood vessels appear inky-black.

A few cases have been reported of central vein closure in young people, especially women, in whom neither circulatory or other systemic change could be found. Coats mentions such a case in a young woman who recovered normal vision. Parsons reported a case in a young woman with irregular menstruation, who recovered with vision of 6/9. I have observed a similar case.

**STEREOGRAM 47. OBSTRUCTION OF CENTRAL VEIN WITH NEURITIS (Hemorrhagic Neuroretinitis).** Unmarried woman, 28 years of age. Menses commonly delayed three or four months. Right eye normal. Left eye vision 20/200. Aside from menstrual disorder nothing abnormal could be discovered by the most exhaustive examinations.

**FUNDUS CONDITIONS.** The optic disc is edematous and slightly swollen. The height of the swelling does not exceed 2 D. (.66 mm.). The

entire retina is edematous and covered with scattered, striate and round hemorrhages. The veins are dilated and tortuous, especially the ascending branches. The arteries are somewhat obscured by retinal edema but otherwise they appear normal. Pressure on the eye empties all veins on the disc with the exception of the ascending temporal. The appearance of the veins on the disc after recovery, indicated that the obstructive pathologic process was anterior to the lamina cribrosa and, probably, involved the temporal veins after they emerged from the nerve. This situation of the obstruction (phlebitis or thrombus) may have caused the nerve to swell. The hemorrhages were gradually absorbed and vision was restored to 20/30 where it has remained for the past eight years. The appearance of this fundus after recovery is shown in Stereogram 49.

**DIAGNOSIS.** In this case the nerve is swollen but not enough to produce hemorrhages. Here, the amount of bleeding and retinal edema is greater than would occur in any obstruction due to mere swelling of the nerve. Pressure on the eye empties the veins indicating an incomplete closure of the central vein of the retina, *i. e.*, a thrombus of the second class.

**ANATOMICAL CHARACTERS.** It has been demonstrated that, in the great majority of cases, the ophthalmoscopic picture of severe hemorrhagic retinitis, already described, is due either to thrombosis or obliterative phlebitis of the central vein, or both combined, very rarely to pressure on the vein (Yamaguchi). Other causes which have been advanced but not universally accepted are: hemorrhagic infarct from closure of the central artery; multiple emboli or thrombi in the retinal veins or arteries; degenerative changes in the retinal vessels and hemorrhage into the substance of the optic nerve.

In order properly to estimate the value of the pathological findings in a given case, knowledge of the methods employed in making the examination is necessary. Formerly, it was the practice to section the globe and optic nerve longitudinally or to cut off the nerve close to the globe and section it transversely. In 1900, Haab pointed out that both these methods were defective, inasmuch as in longitudinal sections an obstruction in a vessel is exceedingly apt to be overlooked, while in nerves cut off from the eyeball, the usual site of a thrombus in the lamina cribrosa was not removed and examined with the nerve. By the present method transverse sections of the nerve and posterior segment of the eye are so made as to include the lamina and, furthermore, every section is microscopically examined, otherwise failure to discover an obstruction in a central vessel

is not conclusive evidence that it does not exist. Michel goes further and does not consider that closure of the central vein can be excluded unless the posterior ends of the central vessels are examined, especially the sharp bend made by them before passing out of the nerve and at their passage through the nerve sheaths, two points which anatomically favor thrombus formation.

Coats says the vein has been found closed at some point in every case presenting the ophthalmoscopic picture of thrombosis in which the optic nerve and lamina cribrosa have been sectioned transversely and every section examined.\* Harms examined 10 cases. In 1 the lumen of the vein was obliterated by endo—and mesophlebitis, and in 6 by an organized thrombus.† While most authorities hold that thrombosis is the usual cause of central vein closure, contrary views have been expressed. Thus, Reimer finds but three cases in ophthalmologic literature which he admits to be true thrombosis. Other cases reported as such he regards as phlebitis or endarteritis proliferans. Verhoff believes that most cases of central vein obstruction are due to primary endovascular proliferation.

Can the ophthalmoscopic picture of retinal thrombosis be caused by closure of the central artery alone? This conception is founded on the well known phenomenon of hemorrhagic infarction, in which the anemic area produced by embolism of an artery becomes filled with extravasated blood. The source of the blood which fills an infarct has been the subject of extensive speculation and research. Virchow suggested all the theories which have since prevailed but did not adopt any of them. In 1860 Cohn proved that hemorrhagic infarct was *not* due to reflux of blood from the veins into the anemic area.‡ Litten, repeating the experiments of Cohn, reached a similar conclusion, *i. e.*, that the blood in hemorrhagic infarction does not come from the veins, but from the capillaries and small collaterals of the surrounding vascular tissue; that hemorrhagic in-

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\*He microscopically examined 16 cases. In 14 the vein was closed by an organized thrombus; in 1 by endophlebitis, and in 1 the findings were inconclusive.

†Of the thrombus cases two were associated with advanced disease of the central artery, three with disease of the vein wall at the site of thrombosis and in one the thrombus formed in an apparently normal vein. In three cases the morbid anatomy was not determined owing to imperfect methods of examination.

‡The following experiments which have been successfully repeated by many investigators, prove that blood does not enter an infarct by backflow from the veins: 1. In a region where tying an artery always causes hemorrhage, if both artery and veins are tied, so that no backflow can take place, bleeding still occurs, in fact, it is greatly increased. 2. In a region where tying an artery is *not* followed by hemorrhage, if both artery and veins are tied, bleeding occurs. 3. If all direct and collateral vessels to a region are tied except the veins, necrosis ensues but no hemorrhage.

farction does not take place in a true terminal vascular system and, "therefore, cannot occur in the retina." These views prevail at the present time. Notwithstanding the experiments of Cohn and Litten, the theory of regurgitant flow from the veins was adopted by Conheim in 1878, a false doctrine, which thus acquired an impetus that has scarcely subsided to this day. Naturally, the theory of hemorrhagic infarction has been applied to cases of severe retinal bleeding in which no obstruction was discovered in the central vein. Opposed to this view are such authorities as Haab, Michel, Coats and Harms, who are convinced that hemorrhagic retinitis does not result from obstruction of the central artery. They admit that in these cases the artery may be obstructed but insist that the vein also will be found closed if the specimen is examined with sufficient care. It is a matter of common observation that closure of either the central artery or its branches is not usually followed by severe hemorrhage. Haab cites, from 21 authors, 39 cases of arterial branch closure in which no trace of bleeding occurred, while only two authors were found who saw faint traces of hemorrhage. On the other hand, in the cases microscopically examined by Wagenmann, Stölting, Knall, Meyerhoff, Reimer and Oatman, hemorrhagic retinitis was attributed to obstruction on the arterial side of the circulation. The majority of these cases were sectioned by the older methods and, therefore, the findings cannot be considered as final. This criticism, however, cannot be applied to either Reimer's or my own case (page 112-113) of hemorrhagic retinitis in which no obstruction was found in the central vein while, in both cases, *nearly* complete closure existed in the central artery. Reimer thinks that, in his case, the course of events was temporary closure of the central artery followed by restoration of circulation, at which time the retinal branches gave way and hemorrhage occurred. This theory would be better sustained if there was any clinical or microscopic evidence that a period of retinal ischemia preceded the bleeding. The mechanism of the hemorrhage in these cases is best explained by the theory of hemorrhagic infarction, which Reimer avoids.

In none of the discussions upon retinal hemorrhage have I seen any reference to the important experiments of Wm. H. Welch, which conclusively demonstrate that hemorrhagic infarction may occur in a terminal vascular system, like the retinal, and that the blood does not necessarily come either from collateral or venous sources. The experiments were made on dogs. All vessels communicating with the intestine were ligated except the main artery and vein. A loop of intestine was then tied above and below so as to shut off all possible sources of collateral



blood. Thus, a terminal vascular system was artificially created. So far, no bleeding occurred. Now, the artery was gradually constricted and repeated measurements showed that hemorrhagic infarction invariably occurred when the blood pressure was thus reduced to  $\frac{1}{4}$  or  $\frac{1}{5}$  of normal. If the artery was further constricted and blood pressure fell below this point, no hemorrhage took place. It was noticed that bleeding always occurred just when gradual compression arrested lateral pulsation (pulse wave) in the artery. During these experiments, at no time, was the circulation entirely arrested. Welch explains the hemorrhage which followed incomplete closure of the artery as follows. The escape of blood, when pressure falls to about  $\frac{1}{4}$  of normal, depends upon certain physiologic properties of blood corpuscles, principally viscosity, by which they stick to the walls of the vessel when not dislodged by the normal pulse wave, and thus time is given them to squeeze out between the endothelial cells. They pass out of the vessel entirely by diapedesis and, as diapedesis requires a certain amount of pressure, no blood escaped when the blood pressure was reduced below a certain minimum. These experiments indicate the possibility of hemorrhagic infarction resulting from incomplete closure of the central artery. Undoubtedly, intraocular pressure tends to prevent hemorrhagic infarction in the retina. It usually occurs where tissue resistance is low, as in the spongy lungs or soft mucosa and sub-mucosa of the intestines. Further experiments by Welch were as follows. Rapid, complete closure of the artery with the vein open was followed by ischemia but no bleeding, which corresponds with clinical experience in closure of the central artery. With the artery open, ligation of the vein was followed by intense congestion and hemorrhage, as occurs in thrombosis of the central vein.

The following explanations have been offered to account for general hemorrhagic retinitis in special cases. Harms is disposed to accept multiple thrombi in the retinal veins, without thrombosis of the central vein, as causative for two of his doubtful cases. Coats admits the possibility of such an event but doubts its occurrence. Alt and Ischeret each found thrombi in the retinal veins but no closure of the central veins. Both Wagenmann and Stölting found multiple thrombi in the retinal arteries with a free lumen in the central vein. Deutschmann diagnosticated, "closure of the central vein from hemorrhage into the substance of the optic nerve," an opinion generally regarded as erroneous.

The central vein like the artery usually is obstructed in, or close behind the rigid lamina cribrosa. Occasionally obstructions form in

other parts of its course. Thrombi have been found by Alt and Wehrli anterior to the disc. In one case by Michel, the thrombus extended from the papilla to the exit of the vein from the nerve and in another it was found 6 mm. behind the lamina. Yamaguci found a thrombus where the vein was compressed as it passed out through a greatly thickened nerve sheath.

Authors agree upon the histologic structure of obstructions found in the central vein, but differ as to their interpretation. The chief difficulty is to differentiate an organized thrombus from endophlebitis. This arises from the fact that after a vein has been closed, either by primary wall disease, thrombosis or both, secondary changes ensue which in a short time present the same microscopic picture, whichever may have been the original cause of occlusion.

**THROMBOSIS.** A thrombus may form in the central vein as a result of local inflammation, manifested microscopically by dense, round celled infiltration of the vessel walls. Such cases are rare and generally occur in the young, especially in those affected with gout, rheumatism, influenza and, probably, in menstrual suppression. In the most common form, which occurs in the aged, there may be proliferation of endothelium but no inflammatory thickening of the walls. Thrombus formation in these cases is ascribed to changes in the endothelial cells and slowing of the blood current, perhaps from obstructive disease in the central artery. Slowing of the current alone is insufficient to produce thrombosis as shown by the following experiment. If, in a living body, two ligatures are carefully applied to a blood vessel, so as not to injure the inner coats, the enclosed blood remains fluid for weeks (Glennard, Baumgarten). Von Recklinghausen regards slowing of the current less productive of thrombus than whirling and eddying of the blood as occurs where a vein is constricted or bent. The so-called marantic thrombus does not result from increased coagulability of the blood, but from lesions in the lining of the vein due to infective phlebitis. Wright, by employing selective stains, usually has been able to demonstrate bacteria in marantic thrombi.

Thrombi form in the central vein by the same processes as elsewhere.\* Coincident with the formation of a thrombus, the mass is in-

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\* (1) *White thrombus*—deposited by the circulating blood and consisting of blood platelets and fibrin. The deposit is quickly invaded by leucocytes. (2) *Red thrombus*, or blood clot, which represents blood suddenly arrested and coagulated *en masse*. This variety consists of red and white corpuscles and fibrillated fibrin. The white corpuscles may be in excess. (3) *Mixed thrombus*, which results from the deposit of white material, by circulating blood, upon the exposed surface of an incomplete red thrombus. (4) *Hyaline*

vaded by phagocytes which proceed to remove the blood elements. In large vessels the further processes of organization are carried on largely by the media, adventitia and especially the vasa vasorum, but in small vessels the size of the central vein, the early stages are carried on almost entirely by the endothelial cells of the vein, which proliferate and replace the thrombus. Connective tissue cells push in from the walls and surrounding tissue and form fibrous septa. Spaces may form in the thrombus, become lined with endothelium and filled with blood by new vessels which enter from the vasa vasorum (Figs. 97-98). From this point the process of organization may pursue either of two courses. In one, the new formed blood channels may enlarge and communication be re-established through the occluded portion of the vein. This is called canalization of a thrombus and has been observed in the central vein by Coats, Harms, Sider—Hugunin and Verhoff\* (Fig. 97). Canalization of a thrombus is exceptional. In the other course, the cellular deposit is replaced by connective tissue and the vein is gradually converted into a fibrous cord, which in course of time, becomes indistinguishable from the surrounding connective tissue (Fig. 92). Under ordinary conditions these processes should be well started within a week, but may be delayed by the presence of pyogenic bacteria, or the walls of the vein may be so degenerated that they cannot furnish new tissue. After a thrombus has been replaced by endothelial cells or fibrous connective tissue, it is often difficult, if not impossible, to determine whether the primary obstruction was a thrombus or the products of endovascular proliferation. In either case the lumen finally is closed by proliferative vasculitis. The appearance of the vessel in front of, and behind the point of closure, may assist in diagnosing the nature of the obstruction. Thus, a thrombus terminates at the point where the first large branch enters the vein† (Fig. 96).

Certain peculiarities in the anatomy of the central retinal vein should be described. In the optic nerve the central artery consists of three layers,

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*thrombus.* This variety occurs in toxic and infectious conditions. It is produced by agglutination (not coagulation) of red corpuscles which undergo hemolysis and form a translucent material which takes Weigert's fibrin stain. These distinctions are recognizable only in thrombi of recent formation, owing to changes produced by the processes of organization which immediately ensue.

\*Verhoff regards the process of canalization as a form of dissecting aneurism.

†Harms differentiates venous obstructions as follows. An *embolus* always excites extensive proliferation of the intima both in front and behind the point of lodgment. In *thrombosis*, on the proximal side, the lumen is narrowed from infiltration and thickening of the walls but there is no proliferation of the intima. On the distal side slight intimal proliferation sometimes occurs. Primary *phlebosclerosis* is diagnosed when high grade inflammatory thickening of the wall exists on both sides of the obstruction.

Fig. 96—A large Collateral Branch joining the Central Vein at the point where a Thrombus terminated.



Fig. 96.



Fig. 97.

Fig. 97—Canalization of a Thrombus, due to the Formation of Coalescing Spaces which become Lined with Endothelium. Walls of Vein Infiltrated with Connective Tissue.

Fig. 98—Another instance of so-called Canalization. In both Figs. 97 and 98, the Vasa Vasorum and Venules of the Optic Nerve are Enlarged.



Fig. 98.







Fig. 99.

Central Vein Closed by Phlebitis and Thrombosis.



Fig. 100.

Sclerosis of Central Vein.  
Walls thickened by proliferation of cells resembling the endothelial.  
(Section 3 m. m. behind globe.)

*i. e.*, adventitia, media and intima. The intima of the artery is distinguished from that of the vein by the presence of a homogeneous elastic membrane, while the vein consists only of an endothelial lining resting upon a thin wall of fibrous tissue. Although the walls of the vein contain no elastic membrane, they do contain a large number of individual elastic fibers. In phlebosclerosis, proliferation of elastic tissue may be the dominant factor in thickening the walls. In such cases the vein is microscopically differentiated from the artery by employing a selective stain for elastic tissue which will exhibit the crenated membrane of the artery as a homogeneous structure which cannot be further resolved by the microscope, while new-formed elastic tissue in a vein consists of individual fibers.

In phlebosclerosis of the central vein, the walls usually are densely infiltrated with cells. Later, the infiltration assumes the form of connective tissue (Fig. 97). In other cases the predominant factor is an exuberant proliferation of cells resembling the endothelial (Fig. 100). These processes may greatly reduce the calibre of the lumen without producing occlusion. In such cases the vein is lined by a smooth, well preserved layer of endothelial cells, over which the blood glides with increased velocity without depositing a thrombus. When, however, this endothelial lining degenerates or is lost, a thrombus forms and the vein is closed by the combined process of phlebitis and thrombosis (Fig. 99). In other cases the vein may be completely closed by cellular proliferation alone. The latter process, undoubtedly, is responsible for closure of the central vein in a large proportion of the cases reported as thrombosis. The following is an example of a vein closed by this method.\*

W. Z. Male, 54 years of age. Had general arteriosclerosis. Left eye. The first ophthalmoscopic examination revealed retinal hemorrhages and neuritis. Four years later he returned to the clinic with intravitreal hemorrhage and high intraocular tension. Recurrent bleeding from the iris and deeper parts continued for the next six weeks when the eye was enucleated. The most interesting pathological feature was found in the condition of the central vein. Commencing about 3 mm. behind the globe, the walls of the vein were infiltrated with epitheloid and round cells which thickened the walls at the expense of the lumen (Fig. 100). Infiltration and proliferation increased towards the eye, gradually encroaching upon the lumen until, in the lamina cribrosa, the lumen was obliterated and the vein replaced by cells and organizing connective tissue (Fig. 101). No

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\*From the Brooklyn Eye and Ear Hospital. Service of Dr. Henry H. Waugh.



blood corpuscles or capillaries existed in the occluding mass. The interpretation of these findings was, endophlebitis proliferans. The process commenced in the intima, the infiltration extending thence to the walls (periphlebitis).

**CLINICAL HISTORY.** Contrary to general experience, Haab, in cases of central vein closure, has met with prodromata in the form of phosphenes and temporary obscurations of vision. Where such a history exists, disease of both central vessels, or increased intraocular tension should be suspected. Almost invariably, one eye only is affected, which illustrates the local character of vascular disease. The few bilateral cases which have been reported arose in the course of acute conditions as in Goh's case of sepsis and Grönow's case in which closure of both central veins was preceded by three days of unconsciousness. In fulminant cases vision may suddenly be abolished or reduced to perception of light. In the majority of cases, however, vision disappears in a spasmodic manner, corresponding to successive hemorrhages. Michel observed that attacks frequently appear on awakening in the morning, in the form of clouds and fog before the eye. In such cases it may be assumed that local conditions favor thrombus formation, which occurs during sleep when the circulation is slow and the blood pressure low. It has been stated that rapid onset and absence of prodromata point to closure of the veins by thrombosis rather than endophlebitis. Location of hemorrhage has an important influence on vision. Thus, in cases of moderate severity vision will be very low if bleeding has occurred in the macula, while in severe cases considerable vision may be present if the macula has escaped. Cases occurring in the course of retinal arteriosclerosis may show temporary improvement in vision from absorption of hemorrhages, but frequently bleeding recurs, the vitreous becomes hazy and total blindness ensues. Sometimes violent glaucoma develops and the eye is enucleated for relief of pain. In incomplete closure a certain amount of vision may be permanently recovered. In very rare cases, usually in the young, vision may be nearly or completely restored after closure of the central vein. After recovery some evidence of previous disease remains, such as slight pallor of the disc, tortuosity of veins, new formed vessels or delicate membranes (Stergm. 49). In the majority of cases, prognosis is bad for the affected eye. As to the fellow eye, almost positive assurance can be given that it will not suffer from the same disease. The presence of white areas in the retina indicates profound nutritive disturbance and they are of serious import as regards vision. The after-history of retinal thrombosis has not been traced in a sufficient

number of cases to assist in estimating the subsequent duration of life. Such prognosis must depend upon the systemic conditions which caused closure of the vein. 'Two of Coats' cases developed symptoms of cerebral thrombosis. He watched five other cases for periods varying from one-and-a-quarter to five years, all of whom remained in good health. Three cases which I have followed for seven, five and three years respectively, are all in good physical condition. In the retinal thrombosis affecting young women with irregular menses the prognosis appears good as regards both vision and duration of life.

COMPLICATIONS. Closure of the central vein not infrequently is followed by inflammatory glaucoma. The percentage of cases in which this occurs has not been ascertained. Its frequency, however, may be inferred from the fact that Coats was able to collect 16 cases within a period of four years. Wilbrand and Saenger give the exact time which elapsed between closure of the vein and onset of glaucoma in 40 cases. Omitting one case in which ten years elapsed, the average time was 14 weeks, varying from two days to fifty-two weeks. I have observed one case of retinitis hemorrhagica followed by iritis without glaucoma.

Cases of thrombosis which first come under observation in a glaucomatous condition, usually escape detection from inability to elicit a reliable history. The following is of diagnostic value in determining the sequence of events. Primary thrombosis of the central vein is nearly always unilateral, while primary glaucoma, nearly always, is a bilateral disease.\* If hemorrhagic retinitis was preceded by the prodromata of glaucoma, namely, temporary obscurations of vision, iridescent halo around artificial lights, premature presbyopia, periorbital pain with redness of the eye, and especially if symptoms of glaucoma exist in the other eye, it may be assumed that glaucoma antedated the hemorrhages. On the other hand, if all these symptoms were absent, and if the glaucoma is limited to the hemorrhagic eye and, furthermore, if loss of vision was rapid and appeared first on waking from sleep, the case may be considered one of primary thrombosis of the central vein with secondary glaucoma. The expulsive hemorrhage which occurs in glaucoma after operations, or even spontaneously, is subretinal and supposed to come from the choroid. The depth of the anterior chamber is of diagnostic importance. Coats has brought out the important point that in glaucoma

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\* v. Graefe regarded the association of retinal hemorrhages and glaucoma as too frequent to be merely coincidental. His observations were made in 1869, before thrombosis of the central vein was described and separated from glaucoma.

following closure of the central vein, the anterior chamber is not, as a rule, shallow. In 12 cases which he microscopically examined and noted the depth of the chamber, it was normal or increased in 11, and rather shallow in one. Therefore, in a given case of monolateral glaucoma, in which loss of vision antedated pain in the eye and the fundus cannot be seen, if the anterior chamber is very shallow, an intraocular growth is suspected. If, however, the chamber is of normal or increased depth, this is evidence in favor of primary thrombosis of the central vein. A deep anterior chamber is of more diagnostic value than a shallow one, inasmuch as in all cases of long standing glaucoma, subject to recurring inflammatory attacks, the area of adhesion between the iris and cornea will gradually increase until the anterior chamber is obliterated, as in the following case.\*

Male, 54 years of age. The right eye had been in a condition of inflammatory glaucoma for several months. There was no perception of light. The loss of vision preceded pain. The anterior chamber was shallow. No view of the fundus could be obtained. Left eye, normal. Evidently, this was a case of glaucoma secondary, either to thrombosis or tumor. Considering the duration of the inflammatory glaucoma, the shallow chamber was not of diagnostic importance, but pointed to intraocular growth. The eye was enucleated and examined. No tumor was found. The optic nerve and posterior segment of the eye were sectioned transversely and every section examined. In the posterior part of the lamina, the central vein was occluded by a well organized thrombus. Behind the thrombus, for a distance of 4 mm., the vein was filled with a disorganized blood clot, which terminated at a point where a large collateral joined the central vein (Fig. 96). The diagnosis was, glaucoma secondary to thrombosis of the central vein.

The mechanism by which closure of the central vein and retinal hemorrhage causes intraocular tension to rise has not been positively determined. Primary glaucoma does not appear to induce thrombosis of the central vein. When these conditions coexist I have found wide spread degeneration of all the retinal vessels. It is inconceivable that interference with the lymph channels around the optic nerve could cause glaucoma, inasmuch as no amount of pressure on the nerve or choking of the disc influences eye tension, so long as anterior drainage is free. Hemorrhage into the vitreous is uncommon in thrombosis. It occurred in none of the 16 cases examined by Coats. Therefore, the glaucoma is not due to ex-

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\*Treated by Dr. G. A. Hare, at the Manhattan Eye, Ear and Throat Hospital.



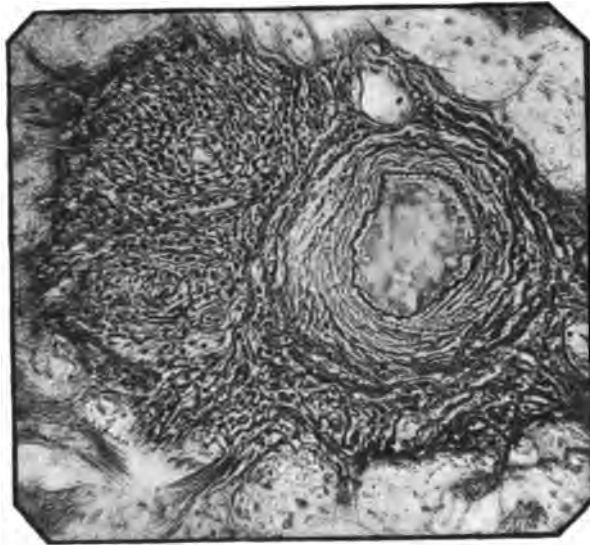


Fig. 101.

Fig. 101—From the same specimen as Fig. 100, but section through the Lamina Cribosa. The Lumen of the Vein (to the left) is Completely Occluded and the Vein Replaced by Cells and Connective Tissue.



Fig. 102.

Fig. 102 — From a case of Glaucoma secondary to Thrombosis of the Central Vein. The Anterior Chamber is deep but the Pectinate is thickened and Fontana's spaces are filled with Pigment and Cells.

travasated blood which has increased the volume of the vitreous. The lens was not disproportionately large in any of Coats' cases. In several cases of thrombosis, glaucoma has been precipitated by the use of a mydriatic, which shows the glaucomatous tendency of this affection.\*

Angiosclerosis alone or combined with thrombosis is the dominant factor in closing the central vein. Sclerosis of the channels which drain the anterior chamber, also appears to be causative in certain cases of glaucoma. Notwithstanding this apparent relationship, sound objections exist against attributing both closure of the vein and a subsequent glaucoma to vascular degeneration. Primary glaucoma, whether due to angiosclerosis or some other cause is nearly always bilateral, while glaucoma following closure of the central vein is unilateral and, moreover, always occurs on the same side as the thrombosis. Therefore, the deduction is that post-thrombotic glaucoma is secondary to, and directly dependent upon closure of the central vein and hemorrhage. The glaucoma appears to depend upon the amount of blood extravasated. Certain, fairly constant peculiarities are observed in the anatomy of post-thrombotic glaucoma which distinguish it from the usual chronic type. (*a*) The anterior chamber is not shallow; (*b*) The fibers of the pectinate ligament frequently are thickened and the spaces of Fontana filled with pigment and endothelium (Fig. 102); (*c*) A sluggish, localized inflammatory process exists in the sinus of the anterior chamber, manifested by slight peripheral adhesions between the iris and cornea, and sometimes by cellular infiltration of the tissues around Schlemm's canal. Recent iritic adhesions were found in 11 of Coats' cases. It is remarkable that in 9 of these the adhesions were limited to the temporal side of the chamber and, when present on both sides, were much narrower on the nasal. This differs from common glaucoma in which the iris is uniformly attached all around.† In thrombotic cases increased tension is not due to slow filtration from increase in the albuminous constituents of the aqueous, inasmuch as the aqueous appears to be less albuminous than in ordinary iridocyclitis. The most reasonable explanation for development of glaucoma after intraocular hemorrhage, has been offered by Baques who, in a case of retinal bleeding found Fontana's spaces filled with leucocytes and proliferating endothelium. There was also cellular infiltration and thickening of the pectinate ligament. This was regarded as a preglaucomatous con-

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\* In four of Coats' cases, glaucoma followed dilatation of the pupil.

† With the possible exception of certain hemorrhagic cases of glaucoma in which bleeding occurred early in the disease.

dition, caused by the presence in the aqueous of irritating chemical products from decomposing blood and dead retinal elements. These products are supposed to produce local infection and inflammation in the pectinate which thickens its fibers and gradually converts its open meshwork into solid tissue. To this should be added blocking of the filtration spaces by phagocytes loaded with products of degeneration from the hemorrhagic areas. This theory is supported by Inouye, who calls attention to the fact that toxins from extravasated blood are hemotactic, *i. e.*, they stimulate the tissues to furnish material for cicatrization. After intra-ocular hemorrhage the pectinate ligament is subjected to this action and responds by proliferation of tissue. If the retinal vessels are sclerosed, absorption of blood will be delayed and the action of the blood toxins prolonged.

The following case which came under my observation appears to illustrate Baques' theory. A laboring man, 58 years of age, with well advanced arteriosclerosis, entered my clinic complaining of blurred vision in the left eye. Right eye presented evidence of retinal arteriosclerosis. Ophthalmoscopic examination of the left eye revealed the picture of complete retinal thrombosis. Three weeks later violent glaucoma, with deep anterior chamber, developed and the eye was enucleated. The appearance of the filtration angle is shown in figure 102. Points of recent adhesions between the root of the iris and the pectinate existed, especially on the temporal side of the eye. The adhesions were situated at a slight distance from the angle of the chamber. The adhesions were readily broken as has occurred in the section shown in the cut. The strands of the pectinate were thickened and Fontana's spaces completely filled by endothelial cells, leucocytes and pigment. Figure 103 shows a section of retina destroyed by hemorrhage from which the extravasated blood has been removed. Pigment-laden leucocytes are seen passing out into the vitreous.

It is not clear why the temporal side of the anterior chamber should be more subject to inflammation than the nasal, unless, as appears true, retinal hemorrhage is always more extensive in the temporal half and the irritating products from decomposing blood are discharged chiefly through this side of the chamber. The relative frequency with which hemorrhage occurs on the two sides of the eye is shown by Ammann, who collected 13 cases of branch retinal thrombosis, all of which occurred on the temporal side.

Iritis is an occasional sequel to retinitis apoplectica. I have seen one case which recovered after resorption of the extravasated blood.

Probably, like postthrombotic glaucoma, it is caused by the local action of toxins from decomposing blood.

**BRANCH CLOSURE.** Closure of a branch retinal vein produces the picture of hemorrhagic retinitis limited to a section of the fundus, drained by the occluded vessel (Fig. 104). If the vein is closed anterior to the disc, the latter probably will be free from swelling. Ammann collected 90 cases of retinal hemorrhage from various causes. Of these, 20 were due to closure of the central vein and 13 to closure of its branches. About two-thirds of the branch cases occurred in the superior temporal and one-third in the inferior temporal branch. Branch closure is said to occur at a somewhat earlier age than closure of the trunk vessel. Arteriosclerosis of both veins and arteries can frequently be seen. Hemorrhage due to closure of small retinal veins occurs in diseases associated with vascular degeneration, as Bright's disease, diabetes, etc.

**STEREOGRAM 48. CLOSURE OF SUPERIOR RETINAL VEIN (Thrombosis).** Right eye of laborer, 49 years of age. Blood pressure 195 mm. Hg. Chronic interstitial nephritis. Left eye is blind from old detachment of the retina. Five days ago the sight in the right eye rapidly failed. Vision now is 20/70. The ophthalmoscopic picture is characteristic of closure of the ascending branch of the central vein. This vessel and its branches are dilated, tortuous and accompanied by hemorrhages. The optic disc is slightly swollen and its temporal margin is obscured. Throughout the region drained by the affected vein, the retina is edematous and opaque. Spots of intense edema or exudation are scattered along the veins, particularly where they make a quick bend and dip into the thickened retina. Both superficial and deep extravasations have occurred. A small, round hemorrhage, situated near the macula, was at first mistaken for a red foveal spot. This error was corrected by directing the patient to gaze into the aperture of the ophthalmoscopic mirror, when the point of fixation (fovea) was found to be above and nearer the disc than the round hemorrhage. The veins not included in the obstruction are somewhat tortuous. This is attributed to retinal arteriosclerosis which is further demonstrated by the descending artery which indents and hides a vein over which it crosses. The mild neuritis may be caused by associated albuminuria or by the situation of the thrombus on the disc.

**DIAGNOSIS.** In this case, the diagnosis of venous branch closure rests upon the presence of hemorrhage, exudation, tortuous veins, comparatively unaltered arteries and retinal edema, limited to the region drained by a branch vein. Primary optic neuritis is excluded on the grounds that so



much edema and hemorrhage could not be caused by moderate swelling of the nerve, and also by the limited area of retina affected.

**STEREOGRAM 49. POSTTHROMBOTIC CHANGES (NEW FORMED VEINS AND MEMBRANES).** Left eyeground of woman, 30 years of age. Two years ago the central vein in this eye was closed. The picture of hemorrhagic retinitis existing at that time is shown in stereogram 47. Vision is now restored to 20/30. The optic disc is physiologically excavated and the cup is filled with new-formed connective tissue which extends beyond the nerve in the form of membranes covering the retinal vessels. The membrane which follows the descending vessels is diaphanous and covered by small vessels, while the ascending vein is enwrapped by a thick, membranous formation which thickens the vessel but does not appear to interfere with circulation. The vascular changes have occurred chiefly on the temporal side of the nerve. The superior temporal vein is obliterated beyond the edge of the disc. Reference to stereogram 47 will show that, in the early period of closure, this vessel appeared to terminate in a deep, round hemorrhage. The macular region to which this vein once was distributed, is now drained by a large number of newly developed vessels connected with the stump of the obliterated vein. The macular region is further supplied and almost encircled by a greatly enlarged branch from the ascending vein.

When, in its passage through the optic nerve, the central vein is closed by a thrombus, circulation is re-established by enlargement of the vasa vasorum and the venules in the nerve (Fig. 98). In a similar manner, if a retinal vein is permanently obstructed, the small tributaries become greatly enlarged and in course of time an efficient collateral flow will be established (Stergm. 30). Another occasional sequel to retinal hemorrhage is the formation of membranes, fibrous bands, etc. (Stergms. 61-62). Why this occurs in one case of bleeding and not in another, is unknown.

**OBSTRUCTION OF BOTH CENTRAL VESSELS.** Clinically, the two conditions, closure of the central artery and of the central vein, are sharply differentiated; but in either affection, microscopical examination frequently reveals more or less obstructive disease in both vessels. In such cases, however, the artery and vein appear never to be affected by the same chronic pathological process, *i. e.*, double thrombosis or double endovascularitis.\* According to Harms nearly complete closure of either the central

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\* Gonin's case of double thrombosis from external pressure on both vein and artery, was a mechanical, not a disease process.

vein or artery, by retarding the blood stream, excites reciprocal disease in the other vessel. In a measure this is due to the fact that the retinal circulation is terminal, and carried on under high (intraocular) pressure. Under these conditions, if circulation is obstructed in either central vessel or in the smaller retinal branches, the entire retinal system is affected. Thus, a thrombus in the central vein, by damming the blood, may excite intima proliferation in the artery. On the other hand, an endarteritic nodule in the central artery may retard the blood current sufficiently to induce thrombus formation in the vein. In a similar manner diffuse sclerosis of the retinal vessels may react upon one or both of the central vessels; in fact, arteriosclerosis not infrequently appears first in the smaller branches. The lumen of a central artery or vein secondarily affected may close first, although obstruction to circulation began in the other vessel. For example, while the ophthalmoscopic picture of either "embolism" or apoplexy shows in which central vessel the disease has terminated, it does not necessarily indicate the one in which it originated. Neither can this point always be determined by microscopic examination.

The following is an illustrative case. The patient was a drunkard, 45 years of age. The surface arteries were highly sclerosed. The day before entering the clinic, vision in the right eye rapidly decreased to movements of the hand. Ophthalmoscopic examination showed the retina covered with hemorrhages, one of which occupied the macula. The retinal veins were tortuous and dilated. Arteriosclerotic changes existed in the retinal arteries. Four months later an extensive hemorrhage occurred into the vitreous. Glaucoma developed and the eye was enucleated. Microscopic examination revealed closure of both central vessels as they passed through the lamina cribrosa, the artery being closed by endarteritis proliferans and the vein by thrombosis. Behind the lamina the vein was almost indistinguishable from the perivascular connective tissue (Fig. 88).

### *Bibliography.*

- Amman, Beitrag. z. prak. Augen. XXXVIII, p. 1.  
 Alt, Am. Jour. Oph. XV, p. 298.  
 Balaban, A. f. A, June 1900; Trans. A. of Oph., XXXIV, 1905, p. 177. A. f. A. XLI, 280.  
 Baumgarten, d. sogan. organiz. d. thromb. Leipzig, 1877.  
 Baques, A. f. O., LXVIII.  
 Bondi, Prag. Med. Wochens., 1901, XXVI.  
 Brücke, British & Foreign Med. & Chirg. Rev., 1857, XIX, p. 183.  
 Clermont, These de Paris, 1900.  
 Coats, O. H. Rep., XIV, 1, 1904, p. 516; XVI, 4, 1906, 516; T. O. Soc., XIV, 1904, 110.  
 Conheim, Untersuch. u. d. emb. processe, Berlin, 1872.  
 Cohn, Klin. d. Emb. gefasskrankh., Berlin, 1860.

- Deutschmann, A. f. O., XXXV, 3, 1879.  
 Glennard, These de Paris, 1875.  
 Goh, A. f. C., XLIII, 1897, p. 166.  
 Goldzeiher, C. f. p. A., XXVIII, 1904, p. 257.  
 Gonin, A. d'Ottal., XXIII, 1903, 2, 219.  
 Groenouw, Graefe u. Saemisch, II, p. 54-55, 625.  
 Haab, Norris & Oliver, IV, p. 515; 28 Versamml. z. Heidelberg, 1890, 210.  
 Harms, Rep. 10th Internal. Cong., A. of Oph. XXXIV, 1905, p. 202. A. f. O. LXI, 1905.  
 Hirschberg, C. f. p. A., X, 1886; XV, 1891.  
 Hormuth, K. m. f. A. Beilag. Festschrift f. Manz u. Sattler, 1903, p. 255.  
 Inoyu, O. H. Rep., XVIII, 1910, 24.  
 Ischeret, A. f. A., 1900, 38.  
 Knapp, A. f. A., 1, 1869.  
 Litten, Über d. Hemorrhag. infark. Berlin, 1879; Zeits. f. Klin. Med. Bd. 1, 1880  
     Arch. f. Oph. LXIII.  
 Michel, A. f. Oph., XXIV, 2, 1878, p. 37; XXII, p. 349; Lehrb. d. Augenheilkunde;  
     A. f. A., XXIV, 2, p. 37; Deutsch. A. f. Klin. Med. XXII, 1878, Zeit. f. Aug. 2, 1889.  
 Moses, Inaug. Dissert., Würzburg, 1896.  
 Parsons, Path. of the Eye, IV, T. O. Soc., XXVII, 1907, 121.  
 Reimer, A. f. A. XXXVIII, 1898, p. 209; 27 Versamml. d. Heid. Oph. Gesell. 1898. A.  
     of Oph. XXIX, 347.  
 Schönewald, Inaug. Dissert., Giessen, 1900, p. 619.  
 Sider-Huginin, A. f. A., LJ, 1904.  
 Stölting, A. f. O., XLIII, 1897.  
 Verhoff, A. of Oph., XXXVI, 1907, p. 1; May 1908, p. 362; A. f. O., LXVI.  
 Von Graefe, A. f. O., XV, 3, 1869, 108.  
 Von Recklinghausen, Deutsch. Arch. f. Klin. Med. 1885, XXXVII, 495.  
 Waggenmann, A. f. O., XXXVIII, 3, 213.  
 Welch, Albutt and Rolleston, VI, 1909, 691 (thrombosis), 762 (embolism). Tr. Assn.  
     Am. Physicians, 1887.  
 Welt, A. f. A., XLI, p. 355; Arch. of Oph. XXX, 1901, 502.  
 Wilbrand u. Saenger, Path. d. Netz. IV, 1909.  
 Würdmann, B. z. A. XXIX, 1898, 100.  
 Yamaguchi, K. m. f. A., XLIII, Beilageheft, 1903, XLI: Neurog. d. Auges, Bd. III,  
     p. 550.  
 Zarhelle, Beitrag. z. Path., ant. u. z. alleg. Path. XLVII, 3, 1910, 539.

## CHAPTER VIII.

### DETACHMENT OF THE RETINA.

In considering the subject of retinal detachment, the layer of pigmented epithelium is regarded as part of the choroid to which it anatomically and pathologically belongs. In its normal position, the retina simply lies in contact with the choroid, the only attachments being at the posterior pole where its fibers unite to form the optic nerve, and at the anterior border where it merges with the subjacent parts to form the ora serrata.\* The retina is held in apposition with the choroid by the vitreous body, the force exerted being equal to that of the intraocular pressure. Separation of the retina is effected in various ways. Thus, it may fall inwards when support is withdrawn, as after extensive loss of vitreous from wounds and operations, or it may be dragged from its bed by contraction of new-formed tissue in the vitreous. In other cases the retina may be pushed away from the choroid by subretinal effusions, displaced vitreous, blood, tumors, cysticerci and penetrating bodies. Detachment often follows the application of blunt force to the eye. In a clinical sense, "detachment of the retina" refers only to that form of retinal displacement in which a translucent fluid accumulates between the retina and choroid. This is known as *serous detachment* and also, in ignorance of its etiology, *simple detachment*.

**SEROUS DETACHMENT.** In this form of detachment separation of the retina usually occurs in an unaccountable manner or from slight causes, as stooping, sneezing, vomiting, etc. Hence, it is sometimes called "spontaneous detachment." As a rule, when it first comes under observation, detachment is extensive and the retina greatly elevated. Exceptionally, the amount of subretinal fluid is slight and the surface of the detachment but little higher than the surrounding fundus, constituting the condition known as *flat detachment*. A slight detachment of the retina may remain "flat." Usually, however, it increases in size. As a rule, it is due to injury, espe-

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\* The terms "detachment" and "reattachment" of the retina are somewhat misleading. Strictly speaking, the retina is not detached, but is displaced or dislocated.

cially in myopic eyes. In such cases a slight blow may be followed weeks later by detachment which at first is flat.

**STEREOGRAM 50. INCIPIENT DETACHMENT OF THE RETINA (Flat Detachment).** Left eyeground of a man, 35 years of age. Two weeks ago, while boxing, he received a stiff blow over the left eye. Ten days later, photopsiæ appeared in the form of fiery sparks and flashes. Vision is reduced and objects appear distorted. The eye is myopic 6.5 D. All parts of the fundus present a red reflex. The optic disc is uniformly reddened and on its temporal side is a narrow, myopic crescent. In the upper temporal quadrant, the surface of the retina is covered by a number of light, parallel lines which disappear on changing the position of the eye. In this region the retinal arteries and veins are darkened and contain no central light streaks. They also are wavy as though following wrinkles in the retina. It cannot be determined by the ophthalmoscope that the surface is elevated. In the lower half of the fundus the retinal vessels are straightened and drawn toward the temporal side, as is common in myopia.

The ophthalmoscopic picture in this case is that of incipient detachment of the retina, the streaked appearance being due to light reflected from inequalities on its surface. It is of the greatest importance to recognize a retinal detachment in the incipient stage. For this reason, myopic eyes which have been injured should be examined frequently for early evidence of retinal separation. While the ophthalmoscopic picture of high retinal detachment is unmistakable, that of flat detachment is recognized with difficulty, in fact, it frequently is overlooked.

**DIAGNOSIS.** In flat detachment, if the subretinal fluid is clear the normal red fundus reflex may not be changed. In such cases the diagnosis of incipient detachment may be established by the presence of some or all the following symptoms:

(a) Functional disturbance consisting of lowered visual acuity, torpor retinæ, metamorphopsiæ, photopsiæ, chromotopsiæ, etc.

(b) Congestion and darkening of the vessels with loss of central light streak and, sometimes, irregularities in their course.

(c) Local hypermetropia demonstrable by the ophthalmoscope.

(d) When underlying choroidal vessels are visible, a parallactic motion between them and the retinal vessels may be obtained, indicating an abnormal difference in levels.

(e) Wrinkles in the retina, demonstrable either by surface light reflexes or by the course of retinal vessels. Not all flat detachments are obscure; some are grey and easily recognized.

Retinal opacity due to closure of a branch artery may be mistaken for flat detachment. Typical cases present the following differences:

(1) The subjective symptoms of detachment are absent in closure of a retinal artery.

(2) In flat retinal detachment, a fair amount of vision is retained, while the area to which an occluded artery is distributed is, or has been, blind.

(3) In arterial closure the surface of the retina is not elevated.

(4) In detachment, the affected region bears no relation to the vessels, but an obstructed artery produces disturbance throughout a triangular area corresponding to its distribution.

(5) The chief distinction is the appearance of the retinal vessels, which in closure are narrowed or obliterated, while in detachment they are abnormally full and dark.

In retinal edema due to choroidal effusion (Stergm. 18) the retina may be separated from the choroid. This condition may be differentiated from simple, serous detachment by the retinal edema which may cover the vessels, and by the presence of other inflammatory choroidal lesions. The possible presence of a young, intraocular tumor must be borne in mind when viewing a circumscribed detachment of the retina.

**SYMPTOMS OF RETINAL DETACHMENT.** The great majority of detached retinas are widely separated from the choroid and occupy one-half, or more, of the fundus (Fig. 105). In uncomplicated serous detachment the eye presents no external signs of irritation. The pupil is moderately dilated and reacts sluggishly to direct light. As a rule, the lens recedes and the anterior chamber deepens. The lens may fail to support the iris and the latter trembles when the eye is moved (iridodonosis). Ocular tension is normal in recent detachment but in old detachment it may be diminished. It is never increased, although in rare cases it may be raised by coexisting disease.\* In high-grade detachment with a wide pupil, it may be possible to see the surface of the retina with the naked eye. With the ophthalmoscope no difficulty is experienced in recognizing a developed detachment. By simple illumination of the fundus a portion of the field will be observed which throws a light colored reflex. Upon detailed examination this is recognized as detachment of the retina by its height and

\* Nordenson examined the intraocular tension in 126 cases of detachment and in only 6 was it increased. Of these, 1 had occlusion of the pupil, 3 had iridocyclitis and in 2 the cause was not apparent. It is doubtful if detachment occurs in glaucomatous eyes, but in an eye with detachment established, conditions might arise in the anterior chamber that would raise the tension.

from a tremulous, wave-like motion which passes over its surface upon the slightest movement of the eye. When first separated from the choroid the retina is transparent and for a brief time presents a red reflex, but it quickly becomes edematous and opaque. Usually, a detached retina has a grey lustre resembling the sheen of a silky fabric. The presence of a very dark green cast, indicates that blood is mixed with the subretinal fluid. Simple detachment seldom forms a single rounded mass, but presents a number of bulging, bladder-like elevations the summits of which reflect light with a whitish sheen (colored plate 4). In other cases the surface is folded, or it may be broken into numerous wrinkles. The border of a detachment may be baggy and overhanging, or it may merge into the fundus with no distinct line of demarcation. Whitish lines or linear folds sometimes are seen at the edge of a detachment, especially along the upper margin of those which recently have changed their position. The detached retina is hyperopic. This fact enables the observer to ascertain the height of the detachment, by measuring with the ophthalmoscope the difference in refraction between its summit and the normal level of the fundus, computing 3 dioptries of refraction as equal to 1 mm. in height. The appearance of the blood vessels in a detached retina is characteristic of the affection. They accurately follow the folds and undulations of the retina, riding prominently over elevations and sinking into depressions. When the retina is detached, the amount of light reflected from the choroid is greatly reduced, consequently, the retinal vessels appear dark or even black. They are also somewhat distended and tortuous from obstructed circulation due to abnormal bends and curves. The tendency of spontaneous detachment is to become complete, in which event the retina is entirely separated except at the nerve and ora serrata. In total detachment if a small vitreous cavity remains, the retina will form a funnel-shaped tube, with bulging walls, at the bottom of which it may be possible to see the optic disc.

Frequently a rent is seen in the detachment through which appears the bright red reflex from the choroid. The laceration usually occurs above, near the periphery, presenting an irregular opening to which may be attached a flap of incurving retina. This rent is supposed to be an important factor in the mechanism of detachment; it certainly is present in a large proportion of all cases although it may be so situated as to escape observation. Any portion of the retina may become detached, but in all cases the subretinal fluid tends to gravitate to the bottom of the eye, consequently, old detachments are situated below. This is the most favorable location,

**COLORED PLATE IV**



**Simple Detachment of the Retina**



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Fig. 103.

Fig. 103—Section of Retina Destroyed by Hemorrhage. The Blood has been Removed and Pigment-laden Leucocytes are seen passing out into the Vitreous.



Fig. 104.

Fig. 104—Closure of a Branch Retinal Vein. Hemorrhagic Retinitis (to the right), limited to the Region Drained by the Occluded Vessel.

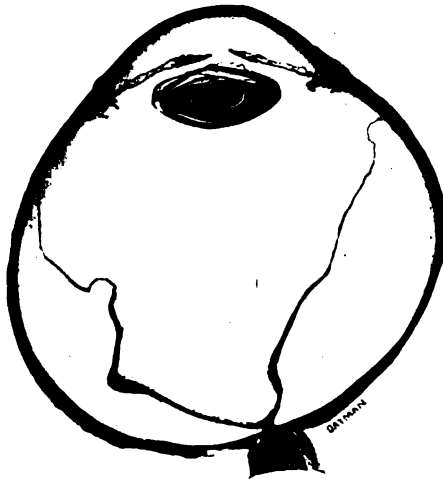


Fig. 105.

Fig. 105—Typical Detachment of the Retina.



Fig. 106.

Fig. 106—Detachment of Retina and Choroid.

as preservation of the upper half of the retina enables the patient to see objects in the lower field. Detachment of the macula is rare. When it occurs the picture may resemble that produced by closure of the central artery, *i. e.*, a bright red macula surrounded by white opaque retina. The red spot is the choroid seen through the thin or open fovea. When hemorrhage or exudation are present in a detached retina, nephritis or other angiopathic disease should be suspected. One case of retinal pigmentation after detachment has been reported.

The symptoms of detachment are not always uniform. The surface tremor, so marked in large flaccid detachments, may scarcely be noticeable in those of moderate size where the retina is tense. Occasionally, detachments remain transparent and fail to exhibit a grey color. Large detachments have been seen in which the retina retained its normal color and fair vision was preserved.\* Such cases raise a suspicion that both choroid and retina were detached (Fig. 106). An authentic case of chorioretinal detachment the borders of which were shelving, is recorded by Spicer. The retina was not opaque but contained clouded areas. The summit of the separated retina was elevated 4 mm. At this level the choroidal vessels were distinctly seen. The visual field was not contracted.

STEREOGRAM 51. DETACHMENT OF THE RETINA. Recent and of high degree. Left fundus oculi of a man 60 years of age. The eye is myopic (-9. D.) Two weeks ago, after violent vomiting, the left eye became nearly blind. At the present time, vision in the upper field of this eye is reduced to perception of light. The pupil is moderately dilated and the anterior chamber deepened. On illuminating the fundus with the ophthalmoscope, the upper half presents a normal red color, but when the patient looks down the reflex becomes light grey. Examination by direct and indirect methods, shows high detachment of the entire lower half of the retina. The detached membrane is thrown into wavy folds which undulate on movement of the eye. Above, where the detachment is shallow, the retina has a reddish cast which gradually merges into a grey color below where the separation between choroid and retina reaches a height of 6 mm. (18. D.). The strong reflection of light from the tops of the retinal folds produces the characteristic "shot silk" appearance. The blood vessels crossing the detachment are very dark and contain no central light streak. As they follow the folds in the retina, their courses become very irregular and apparently interrupted. The optic disc is half enclosed by

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\*Nettleship: Hartridge.

a myopic crescent. Transillumination of the region occupied by the detachment casts no shadow on the pupil.

**DIAGNOSIS.** While no difficulty may be experienced in recognizing a detachment of the retina, its character is not always easy to diagnosticate. It is of vital importance to differentiate simple detachment from detachment secondary to an intraocular tumor. In doubtful cases, a history of injury or the presence of myopia, favors a diagnosis of simple detachment. As a rule, simple detachment occurs rapidly while that due to sarcoma develops slowly. In transillumination, simple detachment offers no obstruction to the passage of light, in which respect it differs from sarcoma which casts a shadow on the pupil. Eye tension may be diminished in simple detachment but is increased or normal in intraocular growths. In simple detachment vision may improve from temporary replacement or change in position of the detachment, while sarcomatous detachment grows progressively worse. Retinal detachment due to tumors may or may not present a characteristic appearance. Sarcoma of the choroid produces a rounded elevation which projects abruptly into the vitreous (Stergms. 74-75-78). Usually, the retina lies in close contact with the summit of the neoplasm or is separated from it by only a thin layer of fluid. The surface of the retina is smooth, free from folds and does not tremble when the eye moves. The color of the sarcoma, which may be white, blackish brown or grey, often is visible through the detachment. In other cases, the layer of fluid between the growth and retina is sufficient in depth to obscure the true cause of detachment. Sarcoma may produce extensive detachment of the retina in regions remote from the tumor. Therefore, in every case of detached retina, search should be made for intraocular growth. Detachment may be due to sarcoma and yet be wholly separated from it (Parsons). This is especially true of small sarcomas situated near the horizon, in which case the detachment may be entirely posterior to the tumor.

Metastatic carcinoma of the choroid produces extensive detachment of the retina at a much earlier stage of growth than does sarcoma. In these cases, the usual situation of choroidal carcinoma at the posterior pole increases the difficulty of diagnosis.

Glioma retinae is situated in the retina itself, differing in this respect from sarcoma and carcinoma which are subretinal growths. Owing to its separation from the sclera, glioma does not cast a shadow in transillumination. Glioma endophytum forms a very evident neoplasm; glioma exophytum, on the other hand, causes early and extensive detachment of the retina.

Conglomerate tubercle of the choroid is prone to invade the retina, and instead of detachment the fundus usually presents the features of an exudative process. Wicherkiewicz, however, has seen extensive detachment of the retina produced by a tubercular nodule. Dor also regards tubercle as a cause of detachment. If, in a given case, tubercle is suspected, the diagnosis would be confirmed if an injection of tuberculin produced reaction in the eye.

Subretinal cysticercus forms a rounded, sharply defined, translucent detachment, beneath which it may be possible to recognize the bluish-grey bladder in which the animal's head appears as a light speck. The parasite may be seen to move within the bladder, but the tremulous motion characteristic of serous detachment is absent.

Cystic degeneration of the retina is given as a cause of detachment. Although large cysts are found in detached retinas, it is doubtful if they ever antedate detachment. These cysts commence with edema of the nuclear layer and the bulging always is outward (Figs. 107-108). Consequently, they cannot be ophthalmoscopically recognized as cysts.

The detached retina of metastatic retinitis is saturated with exudates which render the membrane stiff and immobile so that it does not tremble when the eye is moved. The intraocular tension usually is diminished in metastatic retinitis.

**REATTACHMENT.** In exceptional cases reapposition of a detached retina occurs with more or less restoration of function. Ophthalmoscopic evidence of reapposition usually exists in the form of white or pigmented streaks and spots situated behind the retinal vessels. Such streaks constitute one form of *retinitis striata*. The striæ, which usually correspond to the folds and wrinkles of the detached retina, may radiate from the disc or other points in the fundus. Frequently, they are arranged in parallel lines. In a small proportion of cases the distribution is irregular. These streaks usually represent adhesions between the choroid and retina. A previous detachment, however, cannot be assumed from the mere presence of striæ. Another variety of retinal lines develops in old detachments from stretching and atrophy. In commencing detachment, striæ due to irregular light reflexes from wrinkles in the retina are common (Stergm. 50). A section of retina that has been separated and reattached usually is demarcated from the surrounding fundus by grey or yellowish stripes which may be bordered by pigment. Vitreous membranes have been observed in cases of retinal detachment associated with old hemorrhages. The retinal epithelium beneath a detached retina may undergo ir-

regular depigmentation. In such cases, if reapposition occurs, the retina appears spotted as in choroiditis. In very old detachments the vitreous may contain crystals of cholesterol and produce synchysis. Cholesterol is frequent also in the subretinal fluid. Posterior cataract or phthisis bulbi may ensue in an eye in which the retina has been detached for years.

**OLD DETACHMENT.** In course of time a detached retina becomes highly atrophic and regains its transparency and, if at the same time the subretinal fluid is clear, the detached area may again exhibit a red reflex. The vessels also become lighter in color, although the veins always remain darker than normal.

**STEREOGRAM 52. DETACHMENT OF THE RETINA.** Old, shallow and translucent. Left eyeground of girl, 19 years of age. Stenographer. Refraction normal. Four years ago she was struck in the left eye by a ball, which caused her to faint. Four months later, a positive central scotoma, like a black disc, appeared before the left eye. At that time she experienced flashes of light, which disappeared, but recently have recurred. The scotoma gradually passed away and now, central vision is 20/40. Perception of light is abolished in the upper part of the visual field (Fig. 109). In the left eye the anterior chamber is somewhat deeper than in the right. With ophthalmoscopic illumination the fundus gives a uniform red reflex. With direct examination, the lower half of the retina is seen to be detached. The separation commences on a horizontal line passing just below the disc and increases gradually until at the equator it reaches an elevation of 3 mm. (9. D.). The surface of the detachment is slightly undulating. The color is nearly that of the normal fundus, although faint light reflexes come from the uneven surface. The blood vessels are somewhat dilated and follow the surface inequalities of the retina. The arteries are but little darker than those in the upper part of the fundus, the veins however are nearly black. Just above the upper margin of the detachment are a few horizontal, yellowish streaks, which are interpreted as representing adhesions between choroid and retina in a region formerly detached.

The history indicates that detachment formerly extended above the macula and that the fluid has gravitated below into its present position. That the detached retina is atrophic is shown by its absolute blindness and by its transparency, which permits the passage of light reflected from the choroid. The delayed symptoms may be explained by an unrecognized peripheral detachment which suddenly changed its position.

The prognosis in this case, as regards extension of the process, is good. The detachment was due to traumatism and the cause is no longer



Fig. 107.



Fig. 108.

Figs. 107, 108—Cystic Formation in Retinal Detachment.

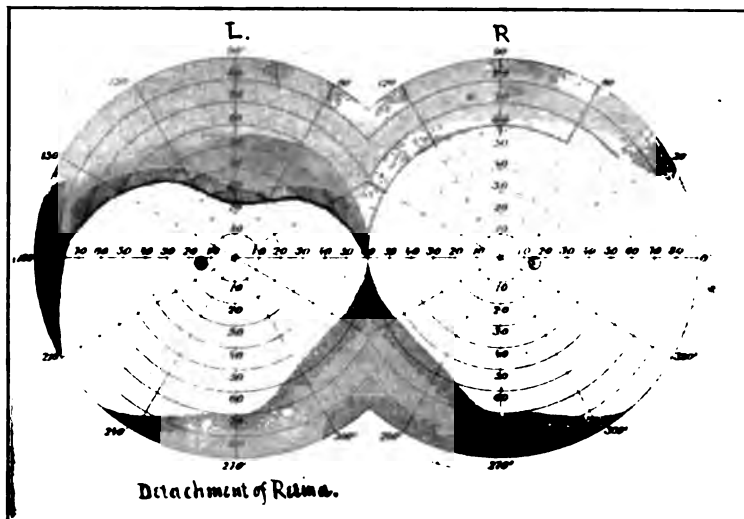


Fig. 109.





active, differing in this from detachment caused by a progressive disease, as occurs in myopia.

**DIAGNOSIS.** Old, low, translucent detachments may be overlooked, owing to their red color, but once discovered they can hardly be mistaken for another condition. The course and dark color of the blood vessels are diagnostic. The form and color of the detachment shown in the stereogram do not suggest the presence of a tumor.

**SYMPTOMS.** Retinal detachment often is preceded by subjective symptoms indicating disturbance in the rod and cone layer. The most common are photopsiæ manifested by a sensation of fiery sparks, darts, balls, etc., passing before the eye. Metamorphopsiæ often are present and objects appear crooked or altered in size. Chromotopsiæ also occur, red, blue, brown and violet visions being the most common. A frequent complaint is of clouds or dust before the eye or of momentary loss of vision.

When these symptoms appear in myopia they should be regarded as signals of impending detachment, although frequently they disappear without such termination. If, however, detachment ensues these phenomena may persist and prove very annoying. Detachment usually occurs suddenly, the event being signalized by the sensation of a cloud or curtain before the eye. Functional disturbance is most noticeable and annoying in rapidly developed cases in which light perception is often retained for months or even years. The undulating motions of the retina cause images which are formed upon the affected region to sway and wave. Reduced central vision usually exists even though the macula is not visibly involved. In some cases, during the forenoon, vision may be good enough to locate objects but later in the day will sink to mere perception of light. The matudinal improvement is due to reposition of the retina which occurs at night while the patient is recumbent. Weeks states that in these cases photopsiæ always are experienced when redetachment occurs. Besides the chromopsia described above, the color sense may be impaired as regards blue and green. Leber thinks this is due to the yellow color of the sub-retinal fluid. As tested by ordinary methods, high detachments are represented in the visual fields as positive scotomata. In case of double detachment below, the fields will show a bilateral upward hemianopsia (Fig. 12). In very flat detachment, in which elevation cannot be demonstrated by the ophthalmoscope, torpor retinæ usually exists, and a field which appears normal in bright light will exhibit a scôtoma when illumination is reduced. Such patients cannot see well on cloudy days. Detachment of the retina occurs most frequently between the 45th and 60th years. It is rare before

the age of thirty. As a rule, one eye only is affected although binocular cases are not unusual. Hortsman reported 106 cases of spontaneous detachment of which 9 occurred in both eyes.

**PROGNOSIS.** The prognosis of retinal detachment in myopia is very bad, but where it is due to Bright's disease or to traumatism, reposition of the retina and restoration of vision may occur. The rods and cones are very resistant to pathological conditions and, if the retina can be replaced within a few weeks or months after detachment, they may resume their function. With the development of atrophy, however, light perception is lost and nothing would be gained were the retina once more in contact with the choroid. Notwithstanding published reports, the permanent cure of a detached retina is a rare event. Although temporary replacement may occur, redetachment ensues in nearly all cases. Uthoff collected all cases of spontaneous retinal detachment in medical literature reported as cured. Of these, 24 per cent had operative treatment, 45 per cent non-operative treatment and 31 per cent no treatment whatever. These statistics render it difficult to estimate the influence of treatment in reducing detachment of the retina, inasmuch as many of those treated might have recovered spontaneously. The best results from treatment are obtained in albuminuria with general anasarca. Uthoff reported a case of nephritis with general retinal detachment in both eyes, in which reattachment followed drainage of the dropsical fluid through the legs by Cruschmann's canulas. A detachment which does not increase in size must be regarded as having terminated favorably.

**ETIOLOGY.** The cause of serous detachment of the retina is unknown despite the frequent opportunities presented for clinical and anatomical study of the affection. Several opposing theories exist, any of which may be true for certain cases but not one of them satisfactorily explains all the features presented by the most common type of spontaneous detachment. The majority of cases occur in myopia. Uthoff found in 500 cases of spontaneous detachment that 60 per cent were myopic. He found also that detachment occurs in 4.5 per cent of all cases of myopia in adults. Statistics showing a lower percentage include children, among whom detachment is extremely rare. Detachment of the retina, however, is not peculiar to myopia but occurs also in both emmetropia and hypermetropia. It is not a rare affection. Schöbl, among 120,000 cases of eye disease, found 280 cases of retinal detachment.

Following are the principal theories which have been advanced to explain the development of simple retinal detachment. They all possess

a logical basis and any of them may be partly or wholly applicable to certain cases.

**EXUDATION OR SECRETION THEORY.** This theory was conceived by Arlt and is supported by Schmidt-Rimpler. The choroid is supposed to furnish a fluid which accumulates behind the retina. The retina is pushed forward against the vitreous, causing the latter to be liquified and absorbed. An associated disease of the ciliary body is supposed to be present which causes further shrinking of the vitreous and increases the retinal detachment. Probably this explanation is true for certain cases of flat detachment; for example, I have seen a young man in whom a low degree of detachment slowly developed in both eyes. The surface of the separated retina was smooth and exhibited no tremor. Such a condition was explained best by the theory of choroidal exudation. There is little doubt that the retinal detachment which occurs in albuminuria is a manifestation of edema in which the transudate comes from the choroidal vessels. In this disease detachment may come and go with the general anasarca as in Uthoff's case described above, or in Raehlmann's in which chemosis of the bulbar conjunctiva and retinal detachment appeared and disappeared together. Raehlmann's case is of further interest as it occurred in a case of nephritis in which there was no retinitis. Galezowski regards rheumatism as a frequent cause of detachment and in such cases he thinks there is an effusion from the choroid analogous to that which is poured into the joints. The exudation theory is particularly applicable to separation of the retina which sometimes develops during the course of nasal sinusitis and orbital abscess and which disappears when the suppurative process is cured. The detachment which accompanies subretinal tumors appears to be caused by a fluid which the choroid exudes in response to the irritating action of the neoplasm. It has been suggested that the detachment which often results from application of blunt force to the eye may be caused by exudation from the choroid. In the exudative stage of choroiditis an effusion often appears between the retina and choroid. This condition frequently is seen in pathological specimens but, clinically, its recognition is uncertain. Stereogram 18 shows an elevated area of the fundus in which it is assumed that the retina is lifted by choroidal exudation. In panophthalmitis the retina is detached by purulent exudates from both retina and choroid (Fig. 32). A subretinal exudate from the choroid is an essential factor in the diffusion theory of Raehlmann; *vide infra*. Cyclitic disease which is mentioned in connection with the exudation theory may yet be shown to exert a greater influence in causing detachment than is generally supposed.

Although the exudative theory holds true for the class of cases described above, strong objections exist to the view that spontaneous, serous detachment is caused by exudation from the choroid. In most cases retinal detachment occurs rapidly or even suddenly, and it is inconceivable that the contents of the globe could suddenly be so greatly increased without raising intraocular pressure. Furthermore, there is no evidence that the choroid is capable of suddenly producing the large quantity of fluid contained in the subretinal space of a large detachment.

**STRETCHING OR MECHANICAL THEORY.** The frequent occurrence of retinal detachment in high axial myopia led von Graefe to the conclusion that separation of the retina in such cases was caused by antero-posterior elongation of the eyeball. This view was supported also by Donders. The mechanism of this theory is as follows. In myopia the posterior segment of the sclera is displaced backward, dragging with it the adherent choroid. The retina, however, is not adherent to the choroid, consequently, slight causes may interrupt its backward excursion and the two membranes may separate. The tendency to separation is increased also by fluidity of the vitreous and hyperemia of the choroid.

This theory is applicable to myopic cases only, but in these it should be given great consideration. Axial myopia predisposes to detachment of the retina, the higher the myopia the greater the predisposition. The full explanation for this may not be forthcoming but, whatever other causes exist, it must be conceded that mere elongation of the globe creates conditions highly favorable to detachment. Thus, the posterior portion of the retina is connected only at the optic nerve, therefore, if this membrane is to follow the choroid and sclera it must be pushed backward by an increase in the volume of the vitreous. The requisite increase of fluid is readily supplied by the normal cells of the ciliary processes, but if these cells are diseased the volume of the vitreous will not be increased and the retina will not be held against the choroid. Myopia tends to produce detachment of the retina by its action on the vitreous. The inert vitreous framework does not increase in size; on the contrary, stretching causes the fibers to rupture and disintegrate, leaving the vitreous in a fluid state. Another result of stretching is thinning and tearing of the retina which renders it easy for a fluid vitreous to filter through and produce detachment.

**HEMORRHAGIC THEORY.** Because of its sudden development, von Graefe once thought that retinal detachment must be due to hemorrhage, but he soon abandoned this view. Of course, hemorrhage from the choroid no longer is advanced as the cause of simple detachment, but it holds





Fig. 110.

**Fibers of the Vitreous, after Abstraction of its Watery Content by the Process of Hardening.**

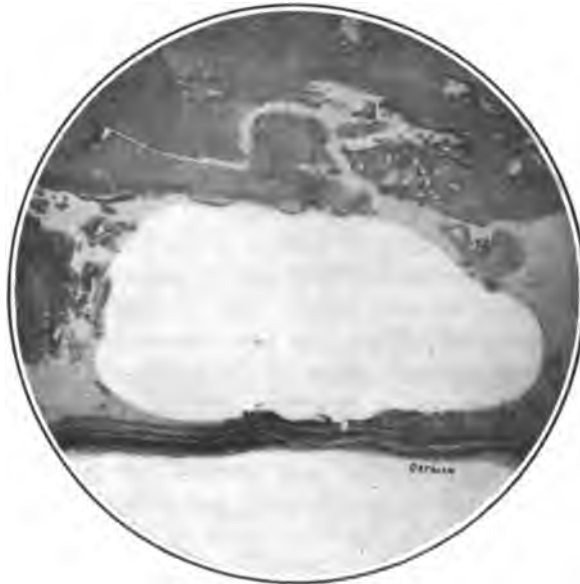


Fig. 111.

**Fig. 111—Large, clear space left in front of the Retina by Contraction of Pathologic Albuminous Vitreous during Process of Hardening.**

good for certain cases of a hemorrhagic type which occur in vascular disease and after injuries or operations (Figs. 115-116).

**DETACHMENT OF THE VITREOUS.** Separation of the vitreous from the retina was first described as a pathologic process by H. Müller. Later, Iwanoff described *localized posterior* detachment of the vitreous, in which a cavity containing fluid is supposed to form between the vitreous and retina. In myopia this cavity is further increased by elongation of the eyeball. The fluid content of the cavity is said to increase until the pressure becomes so great that the fluid filters through the retina and lifts it from the choroid. DeWecker accepted the post vitreous spaces, but thought the presence of fluid forced a hole through the retina.

This theory of the mechanism is faulty. A collection of fluid as conceived by Iwanoff would exert equal pressure in all directions through the soft vitreous, moreover, increased pressure tends to hold the retina in place, not to detach it. Despite the apparent improbability of this theory, the microscopic picture of vitreous detachment, as described by Iwanoff, has repeatedly been confirmed by other investigators and thus has acquired pathologic standing. Nevertheless, it is extremely doubtful that such a condition actually exists *intra vitam*. Greeff, Elschnig and others who have carefully investigated this subject, are emphatic in declaring that the so-called vitreous detachment is an artefact produced by shrinkage when the eye is hardened for microscopic examination. The justice of this criticism is apparent to those who frequently dissect human eyes. The normal vitreous is about 99 per cent water, all of which is abstracted when the eye is hardened, leaving nothing in the vitreous cavity except fibers (Fig. 110). Under pathologic conditions, however, the vitreous contains more or less albumen which forms a visible coagulum in the prepared eye. This albuminous coagulum may contract from behind forward, leaving large clear spaces posteriorly (Fig. 111). Examination of frozen eyes has cast still further doubt upon the existence of vitreous detachment in the living eye. Examination of my own specimens would indicate that the so-called detachment of the vitreous is not present in myopic eyes or eyes with recent detachment of the retina. If, however, in these or other eyes, the vitreous is moderately albuminous, artefacts resembling the vitreous spaces of Iwanoff will be found.



**RETRACTION THEORY (Fibrillary Degeneration of the Vitreous).** This theory, which was presented by Leber and Nordenson, attributes detachment of the retina primarily to a fibrillary degeneration of the vitreous, consisting of an increase in the thickness of the fibers which form its framework. These thickened fibers are supposed to undergo contraction and produce detachment of the vitreous, leaving a space between it and the retina filled with fluid. The final mechanism of detachment is given as follows. Progressive contraction of the vitreous fibers attached to the anterior portion of the retina tear a hole in the latter through which the post-hyaloid fluid passes and detachment is accomplished. By this process the retina may be separated slowly or rapidly without increasing intraocular pressure, because the contents of the eye are not increased, the fluid simply changing position. During all these changes the vitreous fibers remain invisible and the vitreous does not lose its transparency.

Leber and Nordenson's theory, which is widely accepted, is extremely plausible. It is founded upon the most conscientious laboratory work and every microscopic detail described can be confirmed in the human eye and in the eyes of animals in which the retina has been artificially detached. Unfortunately, however, it appears that all the pathologic findings including fibrillary vitreous, vitreous fibers and post-hyaloid spaces, can be found in any eye with a moderately albuminous vitreous, which has been subjected to the laboratory methods of hardening. Especially, is this true in mildly inflammatory conditions in which the fibrin coagulates into a fine network and deposits appear upon the vitreous fibers.

The validity of the retraction theory depends upon proving that the vitreous fibers possess the power of proliferation. The vitreous contains few, if any, fixed cells and, as asserted by Circione, Greeff and others, it is improbable that the vitreous fibers are capable of reproduction, regeneration or enlargement and, furthermore, the only change they ever undergo is to disappear. According to Retzius the vitreous framework tends to break down in adult life and give place to spaces containing a thin fluid, and in the aged the vitreous may be entirely liquified. The condition described in pathological reports as fibrillary degeneration of the vitreous is a normal condition produced in the following manner. The vitreous is a jelly-like body about 99 per cent of which is water. The fluid is enclosed in an extremely delicate membrane and supported by a tangled mesh of long, delicate fibers attached mostly in front. After an eye has been hardened and the water extracted, these fibers are seen streaming out into the vitreous cavity in long tufts like a horse's tail (Retzius) (Fig.

110). They are in no sense pathologic, but if inflammatory conditions exist, exudates and albuminous matter may be deposited on them. Raehlmann denies that the findings of Leber and Nordenson are convincing, inasmuch as the eyes were examined long after detachment occurred and the vitreous changes probably developed after separation of the retina. The retina is not readily drawn from its bed and it is incomprehensible that this can be accomplished by bands in the vitreous which are ophthalmoscopically invisible.\* Highly developed fibers and membranes in the vitreous seldom lead to retinal detachment so long as the ciliary body supplies sufficient fluid to support the vitreous and maintain normal intraocular pressure. Fig. 112 represents one of several specimens which I have examined illustrating this fact. Furthermore, retinal detachment may occur in myopia with no demonstrable change in the vitreous. In the case of ciliary telangiectasis (p. 288-289) the eye was enucleated two weeks after spontaneous detachment. On microscopic examination not the slightest evidence of so-called fibrillary degeneration of the vitreous was present; on the contrary, the vitreous fibers were unusually scanty (Fig. 113). Even shrinking of the vitreous from contraction of inflammatory exudates does not invariably detach the retina (Fig. 112).

The retraction theory requires that the retina be torn, but Nordenson found rents in but 38 per cent of his cases. In the discussion following Leber's presentation of the retraction theory, Samelsohn stated that he had seen a rupture appear on the summit of a detachment coincident with improvement. The detachment which usually follows violent iridocyclitis and shrinking of the vitreous, is a process quite distinct from the fibrillary degeneration and detachment of Leber and Nordenson.

**DIFFUSION THEORY.** Raehlmann affirms that retinal detachment is the result of simple osmosis. To construct this theory he assumes the existence of the following conditions: (1) A highly albuminous exudate from the choroid, situated behind the retina; (2) A faintly albuminous, fluid vitreous in front of the retina. Here are two fluids of different density separated by the thin retina which acts the part of a dialysing membrane. When these two fluids differ greatly in density, diffusion will be rapid; when the difference is slight, diffusion will be slow. In accordance with physical law, the most rapid flow is toward the subretinal space, causing there a great increase of pressure. At the same time, the vitreous cavity,

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\* Greeff truly says, "the cicatricial bands which Deutschmann severs in his operation for the cure of detachment, have never been seen either ophthalmoscopically or microscopically, nor have they been felt with the knife."

having parted with more fluid than it has received, will have its pressure reduced. The high pressure behind the retina is supposed to burst the membrane, causing the rents and fissures seen in detachment, while the low pressure in the vitreous cavity explains the retraction of the lens, deep anterior chamber and diminished tension of old retinal detachment. The frequent absence of lacerations in retinal detachment is urged by Raehlmann as an argument in favor of the diffusion theory and against that of retraction.\*

The diffusion theory is very attractive and meets the requirements better than any other which has been advanced. Nevertheless, it is not free from criticism. It is difficult to accept it for those cases in which detachment occurs almost instantaneously. The theory also shows weakness in elaboration of details. Thus, if diminished eye tension is due to difference in pressure between the vitreous cavity and subretinal space, why does low tension continue when the retina is torn and the two cavities communicate?

That subretinal accumulations of fluid do occur in myopia is shown in Fig. 160. In this eye there was not the slightest tendency for the retina to become detached. Neither did the vitreous contain the usual number of fibrils.

**HYPEREMIC THEORY.** One other theory appears worthy of mention, namely, that of Riva. This author considers that congestion of the choroid alone is sufficient to detach the retina. Although choroidal hyperemia appears inadequate to explain serous detachment, it is a matter of common observation that separation of the retina often follows coughing, vomiting, sneezing, lifting or anything which produces sudden congestion of the head. In these cases it is fair to assume that, when other conditions favor detachment, the determining factor is congestion of the choroid. Congestion may be an important element also in the detachment that follows application of blunt force to the eye. A variation of the hyperemic theory is advanced by Dor, who thinks that in certain diseases the blood pressure is diminished in the ciliary body and increased in the choroid, conditions favorable to retinal detachment.

**IRIDOCYCLITIS.** In this disease it is demonstrable that the retina is dragged inward by contraction of inflammatory bands. Inflammatory exudates are deposited in the anterior part of the vitreous, become vascularized and converted into strands of fibrous tissue attached to the ciliary body and retina (Fig. 114). Contraction of this tissue leads to detachment

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\*Von Graefe suggested pressure of a subretinal exudate as a cause for detachment

Fig. 112—Shrinkage of Vitreous (V) from Inflammatory Exudates without Detachment of the Retina. The Apparent, Slight Detachment is an Artefact.

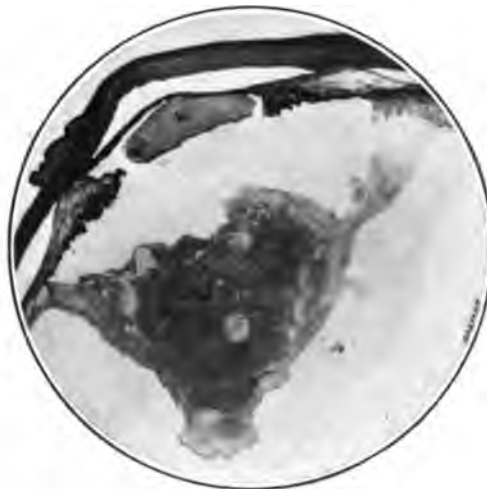


Fig. 112.



Fig. 113.

Fig. 113—Spontaneous Detachment of the Retina. The Vitreous Fibers are Scanty.

Detachment of the Retina due to the Contraction of Bands of Fibrous Tissue Formed by Iridocyclitis.



Fig. 114.





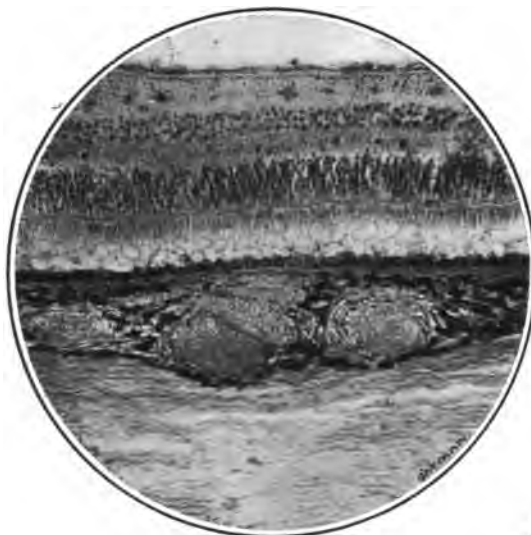


Fig. 115.

Fig. 115—Detachment of the Retina Following Hemorrhage into the Choroid, Probably Due to Thrombosis of the Vena Vorticosæ.

Fig. 116—Choroidal Hemorrhage Causing Retinal Detachment.  
S. C., Sclerotic choroid; B, blood;  
Scl. Sclera.



Fig. 116.



Fig. 117.

Fig. 117—Congenital Detachment of the Retina.

The Retina is Markedly Thickened by Exudates and is Adherent to the Ciliary Body. The Aqueous is Transparent. The Retinal Detachment was Probably Caused by Metastatic Endophthalmitis.

(Specimen of Dr. A. E. Davis.)

of the retina. Progressive softening and shrinking of the globe ensue, constituting the condition known as phthisis bulbi. Probably, the fluid portion of the vitreous is secreted entirely by those cells of the *pars ciliaris retinae* which extend from the ora serrata to the ciliary processes. In cyclitis, these cells suffer and the vitreous is altered in composition and diminished in quantity. In some cases of phthisis bulbi, the presence of adventitious bands attached to the retina is not demonstrable. In such cases it is probable that detachment of the retina is due to the amount of vitreous being insufficient to hold the retina in position.

**HEMORRHAGIC DETACHMENT OF THE RETINA.** This is a clinical classification including only cases of subretinal hemorrhage in which the retina is not ruptured or the vitreous filled with blood. The bleeding may come from either the retina or choroid. Ophthalmoscopically, the detachment has a dark brown or black tint. It occurs usually in the subjects of arteriosclerosis, although Schöbl states that such hemorrhages are seen also in anemic youths and chlorotic girls. I have microscopically examined one such case which occurred in a man, 40 years of age, affected with general arteriosclerosis. The retina was detached by bleeding into the choroid. The source of hemorrhage appeared to be one of the vena vorticosæ which was closed by a fibrous plug, probably an organized thrombus (Figs. 115-116).

**TRAUMATISM.** Detachment frequently follows blows to the eye, brow or temple. Galezowski says that weeks or months may intervene between the blow and resulting detachment. This fact has an important medico-legal bearing. These cases usually are myopic, but the amount of myopia is unimportant, a blow producing detachment with equal readiness in either low or high degrees.

Detachment of the retina may be congenital. J. S. Fernandez has reported an instance in which two out of five children in one family were born with total detachments in both eyes. Syphilis was not established as a cause. From the descriptions, most of the published cases appear to be examples of metastatic retinitis. I have microscopically examined similar congenital cases in which the retina was totally detached and matted together by exudates, while the aqueous and lens were perfectly transparent (Fig. 117).

Certain systemic diseases have been regarded as bearing a causative relation to detachment of the retina. Galezowski, whose experience with this affection is enormous, regards rheumatism as one of the chief causes. Among 1158 cases of spontaneous detachment he found an arthritic di-



athesis in 732. Good reasons appear for attributing some cases to syphilis. Nephritis, undoubtedly is causative. Detachment has been attributed also to diabetes, malaria, erysipelas and infectious diseases, but relationship to these affections remains to be established. A patient of Bellecontre's with Grave's disease, had detachment after division of the cervicle sympathetic. Schirmer reports the case of a girl with slight myopia who suffered detachment of the right retina during chloroform anesthesia.

The various theories which have been discussed in the preceding pages agree in this: that both myopia and fluidity of the vitreous strongly predispose to detachment of the retina. Myopia produces fluidity of the vitreous, but fluidity may develop also in non-myopic eyes. The mechanism of serous detachment appears to consist of a fluid passing from the vitreous cavity into the subretinal space. It is highly probable that this transposition of fluid occurs in various ways. It is easy to understand how it may trickle through an atrophied retina or pour through a rent. Under proper conditions the diffusion theory of Raehlmann, undoubtedly, would be operative. If tangible fibers exist in the retina, as after inflammation or penetrating wounds, their contraction would produce detachment. A more careful study should be made in detachment of those cells which furnish the vitreous fluid. Another class of detachments, not always clinically distinguishable from serous detachment, appear to be caused by exudation from the choroid.

The pathologic anatomy of retinal detachment has been described mostly in connection with its etiology. A few disconnected details, however, remain which require mention. The majority of microscopic examinations have been made in old cases in which degenerative changes in the retina and vitreous may have been secondary to separation (Figs. 118-119).

I have examined one case enucleated two weeks after spontaneous detachment (page 288-289). The vitreous and subretinal fluid contained very little albumin. Vitreous fibers were scanty (Fig. 113). Not the slightest trace of "fibrillary degeneration" of the vitreous was discovered. The detachment was extensive and included the macula. The detached portion of retina was edematous. In the macular region and at the fovea, the rod and cone layer were well preserved (Fig. 120). There was neither exudation from the choroid nor evidence of inflammatory action in any part of the eye.

Saltzman found defects in the lamina elastica of the choroid in myopia. Like all so-called elastic membranes, this layer does not stretch well, and when the eye elongates it becomes fissured. Fluid from the

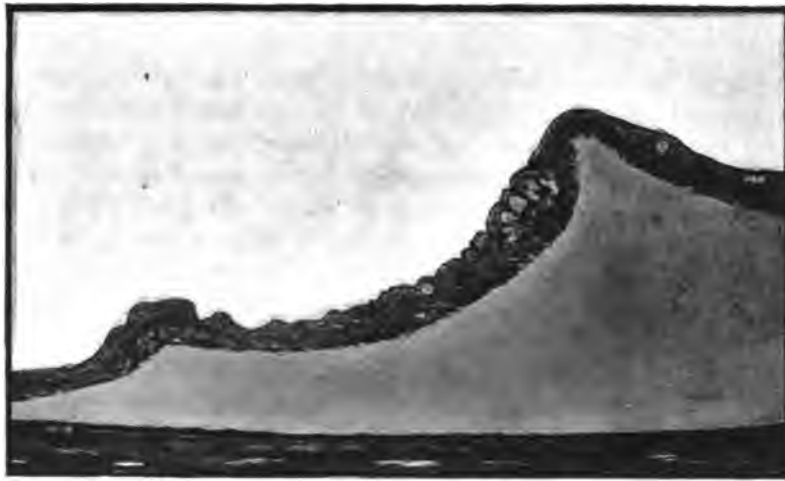


Fig. 118.

Fig. 118—Old, Degenerated Detachment.

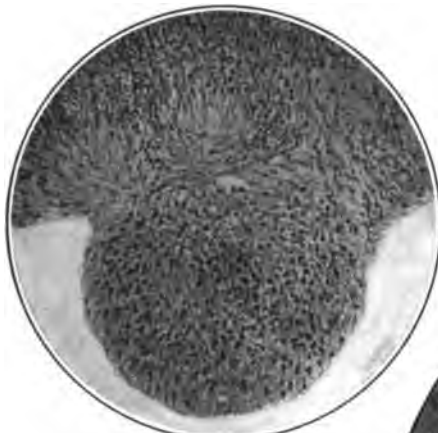


Fig. 119.

Fig. 119—The same, showing Cellular and Connective Tissue Degeneration.

Fig. 120—Retina Two Weeks after Spontaneous Detachment. Section at Macula. Edema of Retina. Rod and Cone Layer Well Preserved.



Fig. 120.



choroid passes through the fissures, causing proliferation of the pigmented epithelium with fusion and scarring of retina and choroid. Choroidal and retinal adhesions tend to prevent detachment, but Hortsman has seen separation occur in such cases. Elschmig considers these points of adhesion as the origin of the rents that occur in detached retinas. The hole is produced by tearing out a piece of adherent retina which remains attached to the choroid when separation occurs. Treacher Collins has microscopically examined two cases in which the exact condition described by Elschmig existed. He also calls attention to the common picture of a retina detached except at one or two places which are regarded as points of adhesion that later may break away, leaving a hole in the retina. Leber considers the inrolling edges of a retinal laceration as evidence of vitreous traction, but the natural tendency of a torn retina is to curl inward. Elschmig saw a case of retinal tear in which inrolling was prevented by a retinal vessel which crossed the opening. Schrenberg examined a case of detachment occurring in the albuminuria of pregnancy. He found extensive exudation between the retina and choroid and thrombosis of many choroidal vessels. Much has been written about exudation from the choroid as a cause of detachment, but the choroid also possesses the power of absorption, and it is possible that removal of subretinal fluid by the choroid may assist in lowering intraocular tension in old serous detachment. Schwalbe describes a lymph space between the pigmented epithelium and the layer of rods and cones which may be of importance in detachment.

In large detachments the subretinal fluid is more albuminous than the vitreous. In a measure, this may be due to its isolated situation where loss of fluid is not easily replenished from the ciliary body.

### *Bibliography.*

- Arlt, *Krankheiten des Auges*, Bd. 3, p. 361.  
 Bellecontre, *Recueil d'Oph.*, XXIII, 1901, 437.  
 Cirincione, *Über d. Entwickl., d. Wirbelthierauges*, Leipzig, 1898; *A. f. A.* Bd. 50, 1904.  
 Collins, Treacher T. *Oph. Soc. U. K.*, XVI, 81.  
 Deutschmann, *Beitrag. z. A.*, 1895.  
 Donders, cited by Schöbl, Norris & Oliver III, 534.  
 Dor, *Mem. d. l. Soc. Med. d. Lyon*, 1904, 173; *Anl. d'Ocl.*, 1901.  
 Elschmig, *Kl. M. f. A.* Bd. XXX, 416.  
 Fernandez, *Arch. of Oph.*, XXXIV, 1905, 338.  
 Galezowski, *Soc. Fran. d'Oph. Nov.* 1892; *Gazette Med. de Paris*, 29; *Rec. d'Oph.* 1884, 1888, 1895, 1889; *Jour. d'Oph.*, 1872.  
 Greeff, *Lehrb. Spec. Path. Anat. (Orth.)* Lief. 11, 2 Theil S. 543. *Arch. of Oph.* XXXIX, 5, p. 515; *A. f. A.*, LIII, 2.  
 Hartridge, *Tr. O. S.*, 1902, 189.  
 Hertel, *A. f. O.*, XLVIII, 2, 1899; *A. f. O.* LVI, 2, 1903.

- Hortsmann, Arch. of O., XXVII, 1898, 475.  
 Iwanoff, A. f. O., XV, 2, S. 1.  
 Leber, Ber. u. d. Heidelberg Oph. Versaml., 1882; Kl. M. F. A. XX., 1882. A. of O. XXXVIII, 1909, 62.  
 Marshall, Deveraux T. O. Soc., XVI, 1896, p. 98.  
 Müller, A. f. O., IV, 1, 372.  
 Nettleship, T. O. Soc., 1885, V, 133.  
 Nordenson, Die Netzhautablösung.  
 Parsons, Oph. Rev., 1905, XXIV, 161.  
 Rachlmann, A. f. O., XXII, 4; A. f. A. XXVII, 1793, p. 1.  
 Rava, Annali di Otolmologie, IX.  
 Retzius, Über d. Baudes Glaskörpers, etc., Bd. 1; Untersuchungen, Bd. VI, 1894, 67.  
 Samelsohn, Bericht u. d. Heidel. Oph. Versaml. 1882; Kl. Med. f. A. 1882.  
 Schirmer, R. Kl. M. f. A., IX, 1871, 246.  
 Schmidt-Rimpler, cited by Schöbl, Norris & Oliver, III, p. 533.  
 Schöbl, Norris & Oliver, III, 543-545.  
 Schrenberg, Kl. M. f. A. 1905, S. 31.  
 Spicer, T. O. S., 1900, 96.  
 Uthoff, Trans. 15 Int. Med. Cong. 1906; A. of Oph. 1906, 535; Heid. Oph. Soc. Sept. 1903; A. of Oph., XXXIII, 1904, 157.  
 Von Graefe, A. f. O. Bd. 1, 361-358; Bericht Kl. Med. f. A. 1886.  
 Von Hippel, Oph. Soc. Heid., Arch. of Oph., XXXVIII, 60.  
 Wagenmann, A. f. O., XV, 209.  
 Weeks, Diseases of the Eye, 1910.  
 Wicherkewicz, Rev. gen. d'Oph., 1900, 546.

## CHAPTER IX.

### DEGENERATIONS OF THE RETINA.

RETINITIS PIGMENTOSA (Pigmentary Degeneration of the Retina). This disease is a non-inflammatory, progressive atrophy of the entire retina, attended by immigration inward of cells and pigment from the layer of tessellated epithelium. The clinical symptoms are remarkably uniform and characteristic, the great majority of cases being typical. The changes observed with the microscope are wholly degenerative. In well developed cases an anterior zone of the retina is covered with a network of jet-black pigment. The pigment deposits consist of small branching lines which form figures that have been compared to bone corpuscles or Haversian canal systems. Generally, the deposits follow the course of retinal vessels or where vessels once existed. The pigment often lies in front of the vessels, proving that it is situated in the inner layers of the retina. Exceptionally, it tends to clump, but in nearly all cases a few branched deposits also are present. The pigment is never connected with white patches, hemorrhages or exudates. Toward the disc the deposits become fewer and fewer until they cease, leaving an unpigmented region around the nerve and macula. In the latest stages of the disease these regions also become pigmented. The amount of pigmentation varies widely in different cases. It may form a close meshwork or be limited to a few scattered, spider-like figures; in rare cases, it may be lacking. The disc assumes the dirty-yellow cast of retinitic atrophy. In old cases scattered colloid dots (drusen) may develop in the retina. As the atrophic process advances, the larger retinal vessels become greatly narrowed and can no longer be traced to the periphery, while the smaller branches become invisible. Shrinking of the vessels proceeds uniformly, differing in this respect from the irregularly distributed degeneration of arteriosclerosis. Rarely do the vessels exhibit white borders or lines, angiosclerotic changes being conspicuously absent. Pigmentation commences in a zone just anterior to the equator in the region where the posterior and recurrent ciliary arteries of the choroid anastomose. From this zone the area of pigmentation extends backward toward the posterior

pole and forward toward the ora serrata. It has been stated that pigmentation is most abundant on the temporal side of the fundus.

Both eyes are affected in all cases. As a rule, the disease is congenital or commences in early childhood. The first and most conspicuous subjective symptom is night blindness (hemeralopia) which exists long before contraction of the field can be demonstrated. The usual history is, that from childhood the patient has seen little or nothing at night or in a dim light, but has had perfect vision in full daylight. This indicates a torpid state of the retina. All perception of light is lost first in the anterior zone of disease, the blind belt slowly widening toward both center and periphery. Vision becomes more and more restricted to the central field, and in course of time, the patient sees objects as though looking through a tube. Orientation suffers, and although the patients may be able to read fine print, they are unable to walk abroad without a guide. Early in the disease, the visual fields may appear normal in a strong light, but when illumination is reduced shrinking of the field and defective vision are shown. In typical cases blindness commences with ring scotoma, although a strong light may be necessary to demonstrate it (Fig. 121). Extension of the scotoma may not be uniform and the result will be crescentic sections of a ring scotoma. In other cases, from the first, the fields present concentric, peripheral contraction (Fig. 122). It will be noticed that both scotoma and pigmentation begin in the same zone and extend in a similar manner, but the amount of pigmentation bears no relation to the degree of retinal atrophy or loss of function. No particular change occurs in the color sense. Sometimes the fields for colors contract before that for white, but in most cases the two contract simultaneously and maintain their normal relations. Photopsiæ are very rare. Leber, among 54 cases of retinitis pigmentosa, saw but one which complained of sparks and flashes of light. The presence of photopsiæ raises the presumption that the case is one of chorioretinitis rather than retinitis pigmentosa. In some, but not all cases, dispersion of retinal pigment leads to exposure of the choroidal vessels. In this disease the choroid is atrophic and the pigment accumulates in the intervascular spaces.

STEREOGRAM 53. RETINITIS PIGMENTOSA. RETINITIC OPTIC ATROPHY.\* Right eyeground of a very dark complexioned woman, 38 years of age. Family history unreliable. Both eyes equally affected. Has never been able to see in a dim light. Concentric contraction of the visual fields

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\* From German Poliklinik. Service of Dr. H. W. Wootton.

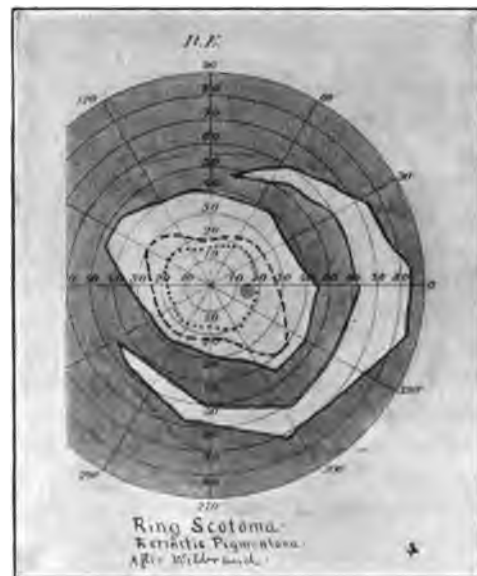


Fig. 121.

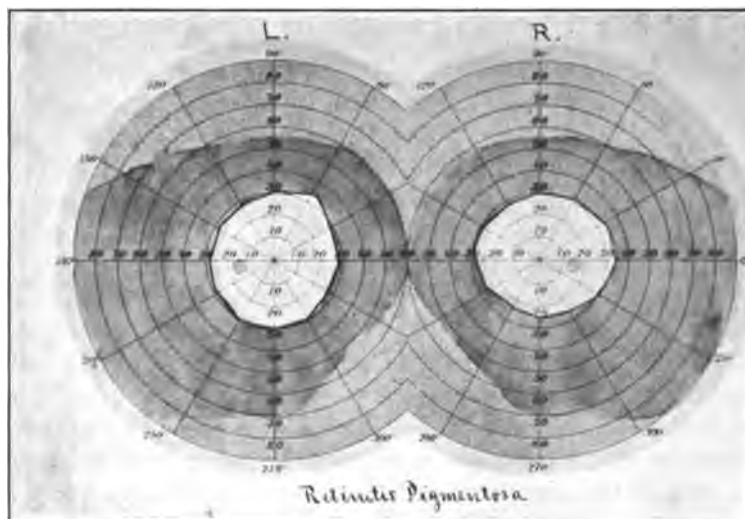


Fig. 122.





progressively advanced, but fair central vision was retained until three years ago when that also failed. At the present time she has bare perception of light. *Ophthalmoscopic Picture.* Probably, this fundus was once very dark. The posterior pole still retains sufficient pigment to hide the choroidal vessels, but in the equatorial and anterior zones the choroid is exposed. Except immediately around the nerve, the retina is covered with large, branching, pigment figures of the peculiar "bone corpuscle" form found in retinitis pigmentosa. The pigment figures lie in front of the retinal vessels when the two are in contact. In addition to the pigment deposits, the retina contains a number of scattered, yellowish dots, supposed to be identical with the colloid bodies or drusen shown in stereogram 27. The main retinal vessels are greatly narrowed, while the smaller branches have wholly disappeared. The optic disc presents the peculiar yellowish-grey discoloration characteristic of atrophy secondary to degeneration of the retina. The disc is rather "hazy" and the markings of the lamina cribrosa are invisible.

**DIAGNOSIS.** The fundus picture is that of retinal atrophy. That this is due to retinitis pigmentosa is indicated by the history of night blindness commencing in childhood and progressive, concentric contraction of the fields, terminating in blindness. Chorioretinitis is excluded by the absence of white fundus patches or other evidence of inflammatory exudation and also by the symmetrical and uniform distribution of the lesions.

**ATYPICAL FORMS. RETINITIS PIGMENTOSA SINE PIGMENTO.** This is a rare form of retinal atrophy with the history and course of retinitis pigmentosa. There is night blindness, contraction of the visual fields, atrophy of the optic nerve and narrowing of the vessels, but the disease is unaccompanied by pigmentation of the fundus.

**RETINITIS PUNCTATA ALBESCENS.** In this form of retinal atrophy the fundus is studded with white dots instead of pigment. See stereogram 54.

**ATROPHIA GYRATA CHOROIDÆ ET RETINÆ.** The above title has been applied to a form of retinal atrophy described by Jacobson and by Colman Cutler but classified and named by Fuchs. It appears to be a disease closely related to retinitis pigmentosa, in which choroidal atrophy is unduly prominent. In addition to all the symptoms of retinitis pigmentosa, there is extreme atrophy of the choroid which commences in the form of round, discreet, white spots distributed throughout the posterior hemisphere. The choroidal spots spread until they become confluent, but before fusion is

complete they are separated from each other by strips and islands of normal choroid. In advanced cases only a narrow ring of normal choroid surrounds the disc, outside of which the choroid is white and atrophic. The retina is pigmented and atrophic, but the choroid is prematurely affected at the posterior pole, differing in this regard from retinitis pigmentosa in which atrophy occurs synchronously in both choroid and retina and a lifetime may elapse before the central field is invaded. The situation and development of the choroidal degeneration suggest an obliterating disease of the posterior ciliary arteries.

A few cases have been reported as retinitis pigmentosa in which pigmentation commenced around the disc and extended toward the periphery.\* All these cases lacked the typical features of retinitis pigmentosa, namely, family history, hemeralopia, concentric contraction of the visual fields, etc. Therefore, they should not be classed as retinitis pigmentosa. The same criticism applies to cases with central, instead of peripheral scotoma. If in genuine retinitis pigmentosa central scotoma is superadded to peripheral scotoma, it is probable that cerebral or optic nerve complications exist.†

Certain types of visual fields may be considered legitimate for retinitis pigmentosa. These are: (1) Peripheral ring scotoma; (2) Concentric contraction; (3) Irregular, concentric contraction; (4) Concentric contraction with retention of peripheral field. Other variations in the visual fields should be accepted as genuine with caution.‡ A few cases have been reported of pigmentary degeneration of the retina that were affected with day blindness (*nyctalopia*) instead of night blindness, the retina being hypersensitive instead of torpid. Such a condition is so at variance with the vast majority of genuine cases that the diagnosis must be regarded as extremely doubtful.

The following form of congenital blindness has been described by Mooren and also by Leber. That the infant is blind is inferred from the presence of nystagmus. During the first six months of life no changes are seen in the fundus. Then occurs thinning of the pigment layer and fine black specks appear in the retina. Later, the fundus presents the picture of retinitis pigmentosa.

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\*Scimemi; Sichel; Knapp; Dujardin.

†Schmidt.

‡Mellinger has recorded a case as retinitis pigmentosa with normal fields, but the case was otherwise atypical. The right eye was blind from glaucoma and the left did not show atrophy of the vessels.

Wilbrand and Saenger quote nine cases reported as unilateral retinitis pigmentosa. This disease is so generally bilateral and its course in the two eyes so symmetrical, that cases reported as confined to one eye should not be accepted as genuine. In considering this and other atypical forms of retinitis pigmentosa, it should be borne in mind that this disease may be simulated so closely by other conditions, especially congenital syphilis, as to be almost if not quite indistinguishable. A doubtful class of cases are those of congenital pigmentation of the retina without impairment of vision.\* It is evident that these could not be cases of retinal atrophy, which is the essential feature of retinitis pigmentosa.

Retinitis pigmentosa is not especially infrequent. Rosenbaum estimated the ratio as 1.5 cases per 1,000 of general eye diseases. Schöbl found ten cases among 11,556 eye patients. The disease exhibits a preference for the male sex. In 191 published cases there were 138 males and 53 females. The Semitic races are said to be more frequently affected than the Aryans.

**COURSE OF THE DISEASE.** In the great majority of cases pigmentary atrophy of the retina appears first in childhood and, undoubtedly, many cases are congenital. The disease develops simultaneously and uniformly in both eyes and its progress is extremely slow. Central vision and color sense usually remain normal in a good light, until the limit of the visual field approaches the 15th parallel, when both fail. This usually occurs between the 40th and 60th years when hopeless blindness develops. Cases are reported in which some vision has been retained until the 80th year. In others blindness has ensued at an early age, sometimes in the 18th or 20th year.

In true retinitis pigmentosa no interruption or arrest of the disease can be expected. Cases in which improvement is said to have occurred are too unusual and remarkable to affect the unfavorable prognosis.† On the other hand, the course of chorioretinitis sometimes may be arrested, and as certain cases of the latter disease are difficult to differentiate from retinitis pigmentosa, individual cases may be treated in the hope that they belong to a spurious type.

**COMPLICATIONS.** Posterior cortical cataract in one or both eyes frequently complicates retinitis pigmentosa. It was present in 54 out of 182 cases cited by Wilbrand and Saenger. These cataracts advance

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\*Juler.

†Scheiss—Gemusus; Groisz.

very slowly or remain stationary. Recent literature would indicate that glaucoma was an infrequent complication of pigmentary atrophy of the retina. About 20 cases are published in which this combination was supposed to exist. In several of these the diagnosis of retinitis pigmentosa was not clearly established. Glaucoma is a common disease and it remains to be proven, that its occurrence with pigmentary atrophy is more than mere coincidence. E. von Hippel examined a case of glaucoma in which the retina was pigmented. The pigment did not come from the retinal epithelium, but was supposed to have separated from the ciliary processes and been carried by leucocytes to the retina. Both Schweigger and Parsons have seen a retina pigmented in this manner and Knape saw a retina which became pigmented after detachment.

ETIOLOGY. The most potent etiologic factor is heredity. Frequently several children of the same parents (siblings) are affected. It follows both by direct and indirect descent. Descent may be intermittent and the disease may reappear after remaining latent for three or four generations. Blood relationship of parents appears to be of etiologic importance, although its influence is denied by some authors. MacNamara affirms that in India where religion successfully prohibits marriage between relatives, retinitis pigmentosa is as frequent as elsewhere. Nevertheless, it is a matter of common observation that in many cases parental consanguinity exists. Both Liebreich and Beyer found it in one half of their cases, but this percentage is too high, subsequent statistics having shown the proportion to be about 25 per cent. The subject of heredity in this and other diseases has been elaborated by Nettleship and he leaves the question undecided, but states that it is of importance in retinitis pigmentosa only when both parents are of tainted stock. Among 976 families, including 1681 persons with retinitis pigmentosa, he found heredity without consanguinity in 23.5 per cent. Consanguinity without heredity in 23 per cent; and combined heredity and consanguinity in 3 per cent. Syphilis has been regarded by many as a common cause of retinitis pigmentosa, and Bocchi states that it usually, if not always, depends upon syphilis of two or three generations back. This view is regarded as erroneous, the error being due to the close resemblance between chorio-retinitis pigmentosa (Stergm. 22) and retinitis pigmentosa. Schön saw seven cases of retinitis pigmentosa of which four were rachitic, truly an exceptional experience. Hemeralopia, with and without pigmentation of the retina, has been observed in connection with chronic diseases of the liver. Landolt considered that an intimate relationship existed between

hepatic cirrhosis and diseases of the choroid and retina, a theory which has not been proven. Pigmentation closely resembling that which occurs in retinitis pigmentosa is often seen in eyes blinded by injuries, especially when the eye contains a foreign body (Fig. 127). Despite the most careful inquiry, a large number of cases remain for which no present or remote cause for the disease can be discovered. This was true in six out of 22 cases reported by Webster, which is about the usual ratio. True pigmentary atrophy of the retina may not be a disease, but a congenital structural defect. This view is strongly supported by its frequent association with imperfections in the nervous system and malformations of the body. The most frequently associated nervous defects are imbecility and idiocy which exist in about 10 per cent of all cases. Deafmutism and deafness are very common. Leibreich in 38 cases of retinitis pigmentosa found 14 deaf mutes, and among 241 deaf mutes he found 14 with retinitis pigmentosa. These percentages are too high, Webster's ratio of 2 cases of deafmutism among 22 of pigmentary atrophy being more accurate. A larger proportion are deaf but not mute. Many cases are associated with epilepsy and arrested body development. Nearly all the ocular malformations have been reported as occurring with retinitis pigmentosa, namely, coloboma of the iris, lens and choroid; microphthalmus, keratoconus, ptosis, high myopia, subluxated lens, persistent hyaloid artery and manifest canal of Cloquet. Colloid masses on the disc (*drusenbildung*) occur in some cases of retinitic pigmentation. Some cases exhibit nystagmus or strabismus. The most frequent coexisting body deformities are polydactylia, harelip and clubbed feet. Other malformations are not unusual. It is possible that retinitis may be a local, premature senility; in other words, the life period of the retina is shorter than that of other body organs. Cases which exhibit vitreous opacities, interstitial keratitis or iritis should be regarded as either syphilitic or tubercular.

**DIAGNOSIS.** In typical cases, the family history, course of disease and ophthalmoscopic picture of retinitis pigmentosa are unmistakable. Atypical and doubtful cases are the exception. Cases which present all features of the disease except pigmentation or those in which white dots occur instead of pigment, are accepted as aberrant types of retinitis pigmentosa. The presence of day blindness instead of night blindness tends to invalidate the diagnosis. The most frequent error in diagnosis occurs in chorio-retinitis with secondary pigmentation of the retina. The formation of pigment spider figures in the retina is not peculiar to retinitis pigmentosa but may occur whenever the tessellated epithelium is displaced or suffers

from lack of nutrition. Förster affirms that syphilis may produce a symptom complex almost indistinguishable from true retinitis pigmentosa. In both diseases there are contracted fields, peripheral pigmentation and yellow atrophy of the nerve. The pigment deposits, however, are not so fine as in pigmentary atrophy and they do not bear any relation to the course of the vessels. Such cases must be uncommon. In syphilis, a careful search usually reveals either deep exudates or white patches. Moreover, the pigmentation is apt to be irregularly distributed, and the visual fields are irregular. In doubtful cases the presence of syphilitic stigmata, *i. e.*, notched incisors, saddle nose, labial scars, etc., would be decisive as would also vitreous opacities, uveitis and interstitial keratitis. In all cases advantage should be taken of serodiagnosis. Pigmentary atrophy may be associated with high myopia, in which event choroidal atrophies due to myopia may legitimately appear in connection with the picture of retinitis pigmentosa.

**ANATOMIC CHARACTER.** The first subjective symptom of retinitis pigmentosa is hemeralopia, and the most conspicuous ophthalmoscopic feature results from dispersion of the pigmented epithelium, all of which indicates profound disturbance in the outer layers of the retina. From the fact that these layers are nourished by the choroid it is assumed that the disease, primarily is an atrophy of the choriocapillaris and that the retinal changes are due to lack of nutrition. This theory is supported by microscopic examinations which show extreme degeneration of the choroid. This satisfactorily explains destruction of the outer retinal layers. The inner retinal layers, however, which include the retinal vessels, are wholly independent of the choroid and something more than disease of the choriocapillaris is required to explain the extreme atrophy which they also undergo. True, an exudative choroiditis may destroy all layers of the retina but, in retinitis pigmentosa, hyperemia and exudation never occur. Neither do the subjective symptoms accord with a primary choroiditis. Thus, night blindness exists for years before ophthalmoscopic or microscopic changes appear in the macula. Ginsberg examined microscopically a case of typical retinitis pigmentosa and found areas of intact choriocapillaris beneath highly atrophic retina and other areas of atrophic choroid covered by intact pigmented epithelium, from which he drew the conclusion that the disease was a primary affection of the neuroepithelial layer. Stock, in a case of central pigmentary degeneration of the retina (not retinitis pigmentosa), obtained the eye for examination an hour and a half after





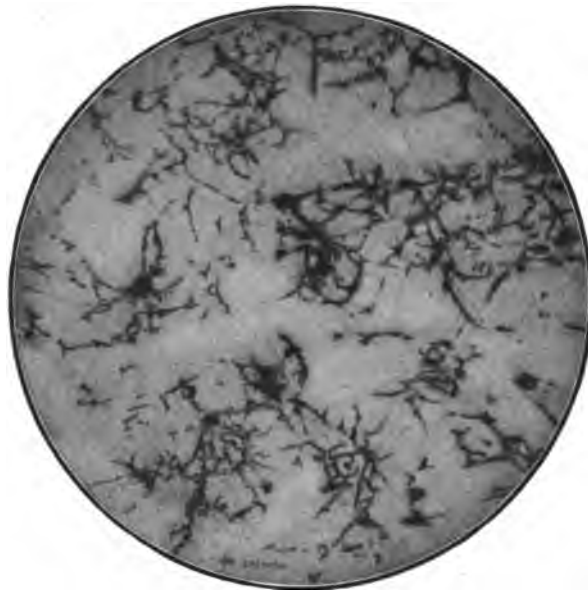


Fig. 123.

Retinitis Pigmentosa. Flat Preparation Showing Bone-corpuscle Arrangement of Pigment.



Fig. 124.

Retinitis Pigmentosa. There is Universal Atrophy of the Retina except in the Macular Region where, as shown in the section, the Different Layers are Distinguishable.

death. The only certain pathologic alteration was degeneration of the rods and cones which Stock regarded as a primary affection. It is difficult to understand how an affection of the neuroepithelium can induce the choroidal atrophy of retinitis pigmentosa. Gonin and Nettleship agree that the cause of the disease may be an insufficient supply of blood to the choroid, connected in some way with the anastomoses between the posterior and recurrent ciliary arteries, which occur in that zone of the fundus where retinitis pigmentosa commences. Experiments upon animals are instructive but indecisive as to the primary seat of retinitis pigmentosa. Thus, section of the optic nerve and posterior ciliary arteries, deprives both choroid and retina of blood and is followed by pigmentation of all layers of the retina. Wagenmann demonstrated that no pigmentation followed section of only the nerve and retinal vessels. Krückmann carefully divided a few posterior ciliary arteries, expecting to shut off the blood from circumscribed regions of the choroid, but migration of pigment occurred only where the external limiting membrane was ruptured. Krückmann's experiments are inconclusive, owing to the free anastomoses between choroidal vessels.

I have had but one opportunity to microscopically examine an eye affected with retinitis pigmentosa. The patient was 64 years of age and totally blind. The left eye was attacked by ulcerative keratitis and enucleated for relief of pain. As the microscopic findings correspond in all essential features with the examinations made by others, it is presented *in lieu* of further description. The anterior section of the eyeball presented the usual features of keratoiritis. The cornea was not perforated externally, but Descemet's membrane was ruptured. The lens, anterior to the equator, appeared normal. At the posterior pole was a zone presenting the usual features of cataractous degeneration. The retina was adherent to the choroid in places and could not be separated without tearing. Flat preparations showed bone corpuscle-shaped deposits of pigment covering all parts of the retina except around the nerve and macula (Fig. 123). Atrophy of the retina was universal, and few normal elements remained, although in the macular region the different layers were still distinguishable (Fig. 124). Here, the nerve fiber and ganglion cell layers (*a*) were thickened and consisted largely of neuroglia and connective tissue. The inner reticular (*b*) and nuclear (*c*) layers were comparatively well preserved. The outer reticulum (*d*) was highly atrophic and the outer nuclei (*e*) were reduced to a single cell layer. The rods and cones were not distinguishable as such but were replaced by fibers

and young connective tissue cells (*f*). The layer of pigmented epithelium (*g*) was irregularly thickened and some cells were loosened but there was no migration of pigment. The choroid was better preserved here than in other regions. In the remaining parts of the retina, the normal structure was replaced by a network of fibrous tissue which took a yellow color with Van Giesen's stain. In most regions this tissue formed a mere tangle. The pigmented epithelium was irregularly distributed and many cells were poorly pigmented (Fig. 125). In some places the pigment had disappeared and in others proliferated. Pigmented cells and free pigment were scattered through the sclerosed retina. The blood vessels were ensheathed in pigment. Their walls were greatly thickened and many were converted into strands of connective tissue. The pigment was deposited first in the perivascular sheaths (Fig. 126) but as the vessels dwindled away it became incorporated in their walls, and in the last stage of atrophy nothing remained of the vessel but a line of pigment. The choroid was highly atrophic, in most places reduced to a single layer of sclerosed vessels and in others to only a layer of pigment. The choroidal pigment was massed against the lamina vitrea, but did not enter the retina. The choriocapillaris had mostly disappeared, but in some places a few small vessels remained even where the retina was highly atrophic.

After severe wounds of the eye or when the eye contains a foreign body, the retina may undergo a form of pigmentary degeneration closely resembling retinitis pigmentosa. An example of this condition is shown in Figure 127. The retinal changes are microscopically indistinguishable from those of retinitis pigmentosa, but the choroid is comparatively normal while in true retinitis pigmentosa it always is degenerated.

In many pathologic reports it is stated that a sharp line of demarcation separates the affected from the unaffected parts. Ginsberg found the macular region nearly normal. Capauner believes the pigmented epithelium may develop ameboid movement, and he regards pigmentation of the retina as due to active emigration of the cells. If the retinal epithelium possesses such function it does not appear to be exercised in retinitis pigmentosa, inasmuch as the cells appear to perish and part with pigment before emigration. Probably, they are passively transported by the retinal currents and deposited in the perivascular sheaths. Possibly, the pigment obstructs the lymph spaces and induces atrophy of the vessels. I have found no pathologic report of a case in which the retina contained no pigment, but in a case examined by Poncet, pigment too fine to be oph-

Fig. 125 — Retinitis Pigmentosa, Complete Degeneration of the Retinal Layers with Irregular Distribution of the Pigment Epithelium.

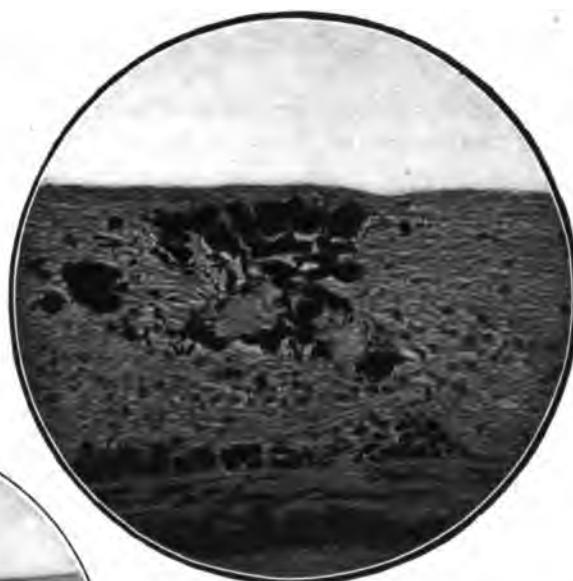


Fig. 125.

Fig. 126—Retinitis Pigmentosa. R, retina; C, choroid; S, sclera. Pigment in the Perivascular Sheaths in the Retina. Choroid Highly Atrophic. Its Pigment does not Invade the Retina.



Fig. 126.

Fig. 127—Retinal Changes due to Injury and Indistinguishable Microscopically from Retinitis Pigmentosa. The Choroid, however, is Normal.



Fig. 127.



thalamoscopically visible was demonstrated with the microscope. The optic atrophy corresponds in degree with that which exists in the retina.

### *Bibliography.*

- Beyer, Inaug. Dissert., 1872.  
 Bocchi, Annali d. Ottal. XXIV, Sup. 21.  
 Capauner, 23 Versaml. d. Heid. Oph. Gesell, 1893, 43.  
 Cutler, Arch. of Oph. XXIV, 1895, 334.  
 Dujardin, Clin. Oph. 1894, 125.  
 Förster, Graefe-Saemisch Handb., VII, Auf. 1, 191.  
 Fuchs, A. of Oph. XXVII, 1898, 484, trans. from XXXII. A. f. O.  
 Ginsberg, K. M. f. A., July 1908; Cent. f. p. A., 1906, 12.  
 Gonin, A. d'O. CXXXV, 101.  
 Groisz, ref. Jahrsber. f. Oph., 1885, 413.  
 Jacobson, K. M. f. A. 1888, 202.  
 Juler, Oph. Rev. 1902, 22.  
 Knape, A. f. A. XLVIII, 1903.  
 Knapp, Trans. Am. Oph. Soc. 1870, 120.  
 Kruckmann, A. f. O. XLVII, 3, 1899; XLVIII, 1-2, 1899.  
 Landolt, A. f. O. XVIII, 1.  
 Leber, A. f. O., XV, 3, S. 1; XVIII, 2, S. 325.  
 Liebreich, Deutsch. Klin. Med., 1861, 6.  
 MacNamara, Dis. of the Eye, 2 edition, 1872.  
 Mellinger, K. M. f. A. XXIX, 171.  
 Mooren, Oph. Beobacht., 1867, S. 260.  
 Nettleship, Tr. O. Soc., XXVII, 1907; XXX, 1910.  
 Parsons, Path. of the Eye, II, 595.  
 Poncet, A. d'Ocul., LXXIV, 234.  
 Rosembaum, Inaug. Dissert., Kiel.  
 Scheiss-Gemus, K. M. f. A., 1875.  
 Schmidt, Inaug. Dissert., Bonn, 1890.  
 Schön, C. f. p. A., 1898, 15.  
 Schweigger, A. f. O., 5, 1, 1859; IX, 1, 1863.  
 Schöbl, Norris & Oliver, III, p. 460.  
 Scimemi, Annal. di Ottal., XXVIII, 69.  
 Sichel, C. f. p. A. Apl. 1877.  
 Stock, Klin. M. f. A., XLVI, 1, 225.  
 Von Hippel, A. f. O., XL, 1, 1894; A. f. O., XLII, 4, 1896; LII, 1, 1901.  
 Wagenmann, A. f. O., XXXVI, 4, 1890.  
 Webster, Tr. Am. Oph. Soc., 1878, 2, 495.  
 Wilbrand u. Saenger, IV. 28; III, 100.

**RETINITIS PUNCTATA ALBESCENS.** Under the above title Mooren described a condition in which the fundus is covered with white dots and the optic disc atrophic. In consequence of Mooren's brief description, the disease was confused with all sorts of punctate conditions of the fundus. Order was brought out of this confusion by Nettleship, Gayet and Fuchs who showed that in addition to white spots in the fundus, a strongly marked group of clinical symptoms usually existed. As now understood, retinitis punctata albescens is a progressive atrophy of the retina presenting all the symptoms of retinitis pigmentosa, except that the fundus is covered with white dots instead of pigment. In Fuchs' case, the smallest dots

were scarcely distinguishable, while the largest did not exceed the diameter of a medium-sized retinal vessel. They were white, sharply outlined, and not bordered by pigment. Some were gathered into groups or rows. The macular region and periphery of the fundus were free from dots.

The disease prevails in certain families, and parental consanguinity frequently exists. The first subjective symptom is night blindness. Concentric progressive contraction of the visual fields commences early. As the disease advances optic atrophy and narrowing of the retinal vessels become apparent. The disease progresses very slowly and some cases appear to remain stationary. The prognosis is much better than in developed pigmentary degeneration. So far as known the etiology and pathology are those of retinitis pigmentosa. Pigment may be present in the retina in specks too small to be visible with the ophthalmoscope as Poncet found in a case which he microscopically examined. The white spots are supposed to be deposited by the retinal epithelium which, instead of parting with pigment, has undergone the peculiar proliferative process by which colloids (drusen) are formed (Stergm. 27). A few of these spots are usually present in advanced cases of retinitis pigmentosa (Stergm. 53).

STEREOGRAM 54. RETINITIS PUNCTATA ALBESCENS.\* Left fundus oculi of a man 36 years of age. Urine normal. Has been unable to see in a dim light since childhood. His father, one brother and two sisters are similarly affected. The fundus conditions are the same in both eyes. The visual fields are concentrically contracted (Fig. 128). In daylight, central vision is 20/30. *Ophthalmoscopic Picture.* Except at the macula, the fundus is covered by groups of small white dots, varying in size from barely visible points to the diameter of a primary retinal vessel. The dots are not bordered by pigment but a few independent groups of fine pigment specks are scattered over the fundus. The optic disc is atrophic. The retinal vessels are uniformly narrowed. The arteries are rather tortuous, but this is regarded as congenital and dependent upon an existing hypermetropia.

Although this retina is not entirely free from pigment the preponderance of the white dots entitles the case to be classed as retinitis punctata albescens rather than retinitis pigmentosa. This case is especially valuable as showing the identity of these two conditions.

DIAGNOSIS. Retinitis punctata albescens requires to be differentiated from the following punctate conditions. Senile colloids and Gunn's dots are limited to the posterior pole or macular region. The lesions of cho-

\* This patient was seen through the courtesy of Dr. H. H. Tyson.

roiditis are never so small as those of punctata albescens. Central punctate diabetic retinitis is distinguished by the size of the spots and presence of hemorrhage. From all the above conditions punctata albescens is differentiated by hemeralopia, contracted fields and ophthalmoscopic appearance of retinal atrophy.

*Bibliography..*

- Gayet, Arch. d'Oph. III, p. 385.  
 Nettleship, Oph. Rev. 1887, p. 181.  
 Fuchs, Arch. f. Augke., XXXIX.  
 Poncet, Arch. d'Ocul., LXXIV, p. 234.

**MACULOCEREBRAL DEGENERATION (Familial).\*** A few peculiar cases have been reported which present a symptom-complex sufficiently uniform and distinct to require independent classification. The one constant, characteristic manifestation is a degenerative process in the retinal macula frequently associated with a similar process in the brain. For descriptive purposes, the affection will be divided into (1), the maculocerebral type, in which both retina and brain are attacked, and (2), the macular type, in which the pathologic changes are limited to the retina.

In the maculocerebral type the patient appears perfectly normal until the second dentition, that is, six or seven years of age, at which time simultaneous failure of vision and intellect ensues. Invariably, both eyes are affected. In developed cases the ophthalmoscopic picture consists of atrophy and pigmentation of the retina in the macular region, bleaching of the optic nerve, and narrowing of the retinal vessels. Small light spots, which do not present the appearance of exudates, may exist in the surrounding retina. Neither retinal hemorrhage nor edema has been described. The earliest ophthalmoscopic change is slight pigment disturbance in the retinal epithelium, which might be regarded as within normal limits. Even this change may be preceded by failure of visual acuity.† The functional eye disturbance consists of central scotoma for green and red, failure of central vision and, sometimes, day blindness. As the disease advances the scotoma for colors increases in size, but does not extend to the periphery. In the late stages the scotoma becomes absolute, and central perception of light is lost. Apparently the degenerative process does not extend to the anterior zone of the retina, and the peripheral field retains its normal boundary, vision and color sense. As central vision disappears, the patient develops eccentric fixation, attended

\*Oatman, American Journal of The Medical Sciences, Aug. 1911; Arch. of Oph., XXXVI, 1907, p. 554.

†Sturgardt.



by nystagmus and lateral deviation of eyes and head. Both nystagmus and lateral deviation may diminish or disappear should the subject become completely imbecile and no longer endeavor to use the eyes. The failure of intellect varies from irritability to imbecility. In some cases epileptiform convulsions ensue. From the few cases reported, it would appear that the retinal atrophy commences in the macula and progresses rapidly for two or three years, after which it extends very slowly. In the brain the disease pursues a similar course, and after the intellect has sunk to the grade of medium imbecility its progress is retarded. No case has become completely blind, but some have become abject imbeciles. In some cases the cerebral degeneration appears to extend to a vital center and cause death.

In the *macular type* of the disease the characteristic degeneration occurs in the macular region, but the intellect is not noticeably affected. It develops later than the maculocerebral type, usually during puberty, when the patient is from fourteen to sixteen years of age. Reports also indicate that in the macular type the pupils are more active, and the nerve does not bleach so early as when the brain is involved. None of the patients were epileptics.

In both types of the disease all the subjects have been reported otherwise in good health and free from malformations. In every instance the affection was confined to one or more members of a childship,\* and not found among ancestors or descendants. Diseases which occur in this manner are termed "familial."† No case has been reported among Hebrews.

The following two cases were typical examples of the maculocerebral type. The childship consists of three children, of whom the eldest and youngest are affected, while the intermediate child, a girl aged ten years, is intelligent and normal in all respects. The parents are not related by blood, but there is an indefinite history of intermarriages among ancestors.

STEREOGRAM 55. MACULOCEREBRAL DEGENERATION. ADVANCED STAGE. Girl aged twelve years. She is the eldest of the three children mentioned above. She possessed good vision and was an excellent scholar until the age of seven years, when vision and intellect began to fail. At the age of nine years she developed epileptiform seizures which have continued at irregular intervals.

PRESENT CONDITION. Large, robust girl. Repeated Wassermann

\*Childship: All the children of the same father and mother.

†Familial disease: One that occurs among members of a childship but has shown little or no tendency to affect ancestors or descendants.



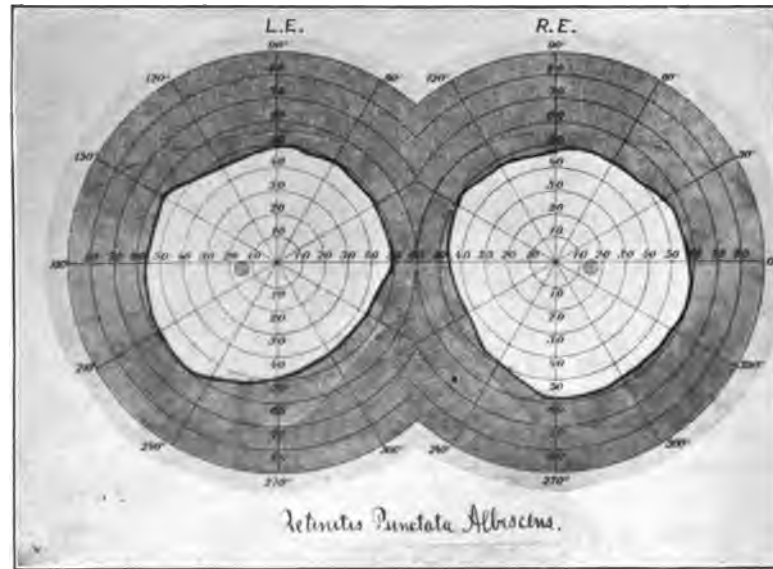


Fig. 128.

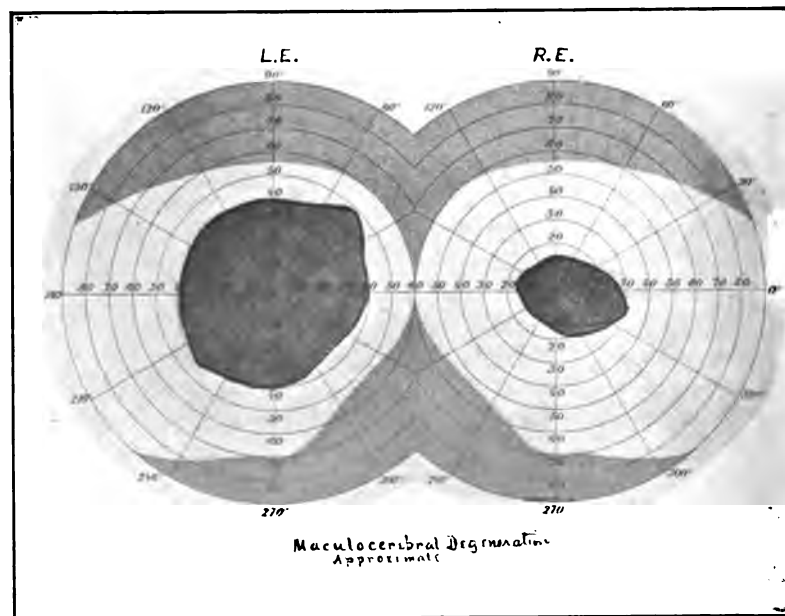


Fig. 129.

tests for syphilis and von Pirquet's test for tuberculosis were negative; no malformations; is a medium imbecile but talks well, and has a good memory for remote events; habits cleanly; myopic (-2. D.); pupils sluggish but otherwise normal; has eccentric fixation, with head and eyes turned to right; lateral nystagmus; has neither night nor day blindness. There is absolute central scotoma in both eyes, roughly estimated as 40° in the right and 10° in the left (Fig. 129). Peripheral fields are not contracted, and possess good perception of white and colors.

**OPHTHALMOSCOPIC PICTURE.** Media clear. Strongly marked tigered brunette fundus. Optic nerve decidedly atrophic. Retinal vessels narrowed but not otherwise changed. The smaller vascular twigs have mostly disappeared. The macular region exhibits a pigmentary atrophy of the retina limited to an area with a diameter about two and a half times that of the optic disc. In this region the fundus is mottled, has a dirty, yellowish-grey color and is peppered with granular and dust-like pigment. A number of ill-defined, light radiating lines, extend outward from the temporal side of the nerve. They appear to be light reflexes. Both eyes are affected symmetrically. In the right eye the retina is more atrophic, the fundus lighter in color and the pigment deposits are being dissipated.

Four years later: The patient has almost reached the stage of complete imbecility. Memory poor. Saliva collects in pharynx and interferes with speech. Peripheral fields not contracted. Can distinguish red in large mass. Central blind area does not appear to exceed 40°. The retinal pigment as far as the equator is greatly thinned and the whole fundus is much paler than four years ago. The central area is veiled as though the retina were converted into a fibrinous membrane. In the macular region the pigment has entirely disappeared and several orange colored choroidal vessels are exposed.

One year later this patient died with symptoms indicating that the degenerative process had extended to some vital cerebral center.\*

**STEREOGRAM 56. MACULOCEREBRAL DEGENERATION; EARLY STAGE.** Right eyeground of boy, aged eight years. Brother of case (Stergm. 55) described above. Wassermann reaction for syphilis and von Pirquet for tuberculosis, negative. Was considered a normal child until six years of age, when failure was noticed in vision and intellect. He is in excellent health; no paralysis; no malformations. Vision in right eye

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\*Unfortunately, I was unable to secure an autopsy.

is 10/200; in left 8/200. Small central scotoma for green and red; peripheral fields normal for white and colors; central fixation; talks well and memory good.

**OPHTHALMOSCOPIC PICTURE.** Media clear. The macula is encircled by a transversely oblong ring of granular pigment, measuring about one disc diameter in length. The enclosed area has a dirty, yellowish cast and contains dark spots which can be resolved into fine black granules. The surrounding retina is covered with dust-like pigment. The optic nerve is white on the temporal side. Retinal vessels, narrowed.

The left eye presents the same picture as the right, except that the macula within the pigment ring is more atrophic and the fundus as a whole, somewhat lighter in color.

Four years later. The disease has steadily advanced. No paralysis. Central scotoma is now absolute. Peripheral fields appear normal. Memory poor. Is apathetic and makes little effort to see. Eccentric fixation with eyes upward. Retina is becoming depigmented. Has had five epileptiform convulsions, the first of which occurred three years ago.

The intermediate child is now fourteen years of age. She is a normal, healthy girl, with perfect visual acuity.

Cases of maculocerebral type were reported first by F. E. Battin of London, in 1903 as "Cerebral Degeneration, with Symmetrical Changes in the Macula in Two Members of a Family." Their vision and mental condition gradually deteriorated, and they were removed to an asylum where eventually they died. They did not become entirely blind.

M. S. Mayou in 1904 reported three similar cases in a childship of seven children. Parents were first cousins. In her first pregnancy the mother miscarried at three months. The second, third and fourth children were born healthy but developed maculocerebral degeneration, while the fifth, sixth and seventh were both healthy and intelligent.

Six years later Mr. Mayou courteously writes as follows: "My cases are still alive and the eye condition remains stationary. The eldest child has become a complete imbecile, and the other two children are in schools for the mentally defective. The mental condition appears to have a slowly progressive tendency, but the children are not blind."

Other cases, supposed to be maculocerebral degeneration have been recorded by Nettleship, and Hirschberg.

**MACULAR DEGENERATION.** As stated, the macular type presents the history and every clinical feature of the disease, except failure of intellect. The period of development, however, is delayed until the age of puberty.

Two examples described by R. D. Battin, in 1897, appear to be the first of either type reported.

Under the title of "Family Progressive Degeneration in the Macular Region of the Eye," K. Stargardt has published the history of two childships which are typical examples of the macular type. The first childship consisted of four children, all of whom were affected. There was no family history of eye disease, consanguinity of parents or syphilis. Aside from the eye affection, the patients were healthy, free from malformations and of average intelligence.

Stargardt's second childship consisted of five children, three of whom developed macular degeneration. Except the macular disease, every member of the childship was intelligent, healthy and free from malformations.

Stargardt cites a further case from Leber's records which appears to be an example of the macular type.

**PATHOLOGIC ANATOMY.** In the absence of a microscopic examination the histologic changes can be surmised only from the ophthalmoscopic picture. This indicates an atrophic degeneration of the retina, commencing in the outer layers. The earliest visible change is loss of foveal reflex and rarefaction of pigment. The loosened pigment is next distributed throughout the retina as a fine dust or specks which exhibit little tendency to wander inward over the retinal vessels. The pigment does not proliferate, neither is it fixed, inasmuch as it eventually disappears. Ultimately, the retina is converted into a thin cicatricial membrane, constituting the delicate film which covers the macular region in advanced cases. When depigmentation is complete, the choroidal vessels are exposed. In my case, in which the choroid was thus revealed, the interspaces contained no pigment, although normally this was a strongly "tessellated" fundus. Depigmentation of the fundus oculi may be due either to local action of some adventitious substance, or the pigment granules may be removed by leucocytes.\* The absence of all evidence of inflammatory action suggests that the retinal atrophy results from nutritive disturbance. Inasmuch as the retinal layer first affected is nourished entirely by the choroid, it might be inferred that the primary lesion is in the choriocapillaris. Furthermore, limitation of the pathologic changes to the posterior pole of the eye corresponds to the distribution of a choroidal vascular circuit. Extreme interest attaches to the work of Stock (*vide infra*). In three cases of primary retinal degeneration he appears to have demonstrated by microscopic examinations that

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\*I have seen complete depigmentation of the eyeground in a diabetic (Arch. of Oph., 1910, XXXIX, p. 392.) (Stergm. 26.)

the primary change was a degeneration of the rods and cones, probably caused by selective action of some toxin in the blood.\* No case reported indicates that the deep layers of the choroid were affected. The light spots described in many cases should be regarded as degenerative rather than exudative. They may have been due either to exposure and thickening of the lamina vitrea or to condensation and thickening of the retinal neuroglia. In Stargardt's, and also in my cases, the spots were not prominent and could be seen with the ophthalmoscope only by the direct method. Their tendency to fade as the light was moved indicated reflexes rather than actual deposits. Probably the optic atrophy is secondary to retinal degeneration, although, as regards cerebral cases, this is not certain. In my second case the nerve was white and vessels narrowed, early in the disease. In the macular type, however, a late secondary atrophy is described.

**ETIOLOGY.** The records throw little light upon the cause of the affection. In one childship congenital syphilis was surmised, but not proved. Certainly, it did not exist in my cases. Racial predisposition, as in amaurotic family idiocy, may be excluded, as none of the cases occurred among Hebrews, but were distributed among various other races. Hereditary tendency to eye diseases did not prevail in any of the affected families. Blood relationship of parents may be measurably causative; thus, it existed in four of the six childships affected with the maculocerebral type, and in one of the four childships affected with the macular type. The fundus changes are not unlike those which occur in the aged, as though the retina were affected with a local, premature senility. Nettleship suggested that a cause for retinal degeneration in children might be found in nutritive disturbance due to the exanthemata or other exhausting conditions. In the pigmentary atrophies of the retina, Stock argues that an autocytoxin exists in the blood, which exerts a selective action upon the rods and cones. Stargardt, applying this theory to his cases, suggests a toxin with a selective action for cones only. Dr. Max Schlapp, who observed my cases also regarded the pathologic changes as due to the action of an autotoxin upon the nervous elements of the brain and retina. It is important to note that the maculocerebral type appears with second dentition, while the macular type is delayed until the period of puberty. It is improbable that this is mere coincidence. More likely the developmental forces active during dentition and sexual evolution are here perverted to stimulate a latent degenerative tendency.

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\*Possibly syphilis.

**DIFFERENTIATION.** Several affections exist which, while differing from each other, all bear some resemblance to maculocerebral degeneration. These are, hereditary macular anomaly, retinitis pigmentosa, amaurotic family idiocy, central chorioretinitis, and certain unclassified cases.

**HEREDITARY MACULAR ANOMALY.** This condition has been well presented by F. Best, who described the remarkable ophthalmologic history of a family of whom 8 members presented a defect at the macula. In all other respects, however, the cases differed essentially from maculocerebral degeneration. The family was traced through five generations, and 59 individuals were examined, of whom 31 presented some affection of the eyes.\* The 8 affected with macular anomalies were distributed among 5 childships. The defect occurred by direct, indirect and collateral inheritance.† In 6 of the cases both eyes were affected, and in 2 the change was rudimentary and limited to one eye. In every case the defect was situated immediately below the point of fixation, a uniformity which Best regarded as a remarkable example of persistence in hereditary transmission. Among the cases with macular defect the following additional abnormalities were found: high hypermetropia, astigmatism, strabismus, persistent pupillary membrane, and one case of total color blindness. None of the cases had epilepsy, imbecility, or optic atrophy. Aside from the one case of total color blindness, the color sense was normal in all. Visual acuity was blunted, but none of the subjects was incapacitated. The poor vision had existed since early childhood, and was non-progressive. During the eight years they were under observation no changes occurred in the eyes. From these facts Best justly decided that the condition was an hereditary macular anomaly.

Maculocerebral degeneration differs from such congenital defects in its family history, late development, progressive course, cerebral complications, central color scotoma, optic atrophy and invariable bilateral occurrence.

**RETINITIS PIGMENTOSA.** At first glance, maculocerebral degeneration appears like an atypical retinitis pigmentosa. Both affections develop in early life, are slowly progressive, and present a symmetrical picture of

\*Macular defects, 8: opaque nerve fibres, 1: one-sided amblyopia, 5: double amblyopia with nystagmus, 2: strabismus convergence, 3: pigment points in eyeground, 6: senile cataracts, 5: punctiform opacities at posterior pole, 6: ptosis and telangiectasis of lid, 1: glioma, 1: hypermetropia, 7: hyperopic astigmatism, 8: anisometropia, 7. None of the cases were myopic.

†Direct inheritance: From parent to child. Indirect inheritance: From grandparents, uncles, or aunts. Collateral inheritance: Occurrence of the same anomaly among brothers and sisters; thus, familial disease occurs by collateral inheritance.



atrophy in each retina. Furthermore, it is estimated that 10 to 13 per cent of retinitis pigmentosa cases are idiots. These analogies, however, are more than counterbalanced by the following features in which the two diseases differ. The most constant symptom in retinitis pigmentosa is night blindness. Supposititious cases, in which day blindness existed, are too few in number to vitiate the rule. The presence of night blindness more than anything else has caused retinitis punctata albescens, retinitis pigmentosa sine pigmento, and gyrate atrophy to be classed as aberrant forms of retinitis pigmentosa. Opposed to this are 20 cases of maculocerebral degeneration, none of which had hemeralopia, while 3 saw better in a dim light. Retinitis pigmentosa is eminently hereditary and the persistence with which it reappears in families renders it easy to trace its history. In maculocerebral degeneration, however, the family trees are remarkably exempt from eye diseases. True retinitis pigmentosa invariably commences near the periphery and all parts of the retina are invaded, the macula last of all.\* Macular degeneration, on the contrary, commences in the macula, and does not appear to extend into the periphery. In retinitis pigmentosa the color sense fails only with light perception, but in maculocerebral degeneration one of the earliest manifestations is central scotoma for green and red. The idiocy associated with retinitis pigmentosa always is congenital, while the subjects of maculocerebral degeneration are born with normal intellects, which subsequently may be destroyed by the disease. Malformations of all kinds, so common in retinitis pigmentosa, are lacking in maculocerebral degeneration.† Finally, retinitis pigmentosa is transmitted for generations, while maculocerebral degeneration is a familial disease.

**AMAUROTIC FAMILY IDIOCY.** Hirschberg remarked that his case presented some symptoms not unlike amaurotic idiocy, but differed in other and essential points. Nettleship, however, is disposed to trace a relationship between the two diseases. He suggests that amaurotic family idiocy may sometimes pursue a mild course and permit the child to grow up, and cited the cases of J. D. Battin and Mayou as possible examples. He avoided the racial difficulty on the supposition that Jewish blood flows in many gentiles. Apparently the two diseases are entirely distinct. Amaurotic idiocy appears to occur exclusively among Hebrews. Cases reported among other races have not been accepted as genuine. The lesion is a primary degeneration of the ganglion cells throughout the nervous system in-

\*Cases of Knapp, Dujardin, Scimmi and Sichel, reported as retinitis pigmentosa commencing at the macula, are lacking in other essential features of that disease and cannot be accepted as genuine.

†Hirschberg's cases presented stigmata, not deformities.

cluding those of the retina. The disease develops a few months after birth and eventuates in general muscular paresis, idiocy, and blindness, the patient usually dying within two or three years. In contradistinction to this history, maculocerebral degeneration develops in late childhood, and, so far, only among gentiles. In no case has muscular weakness, paralysis, or ill health been attributed to the disease; neither has any case become entirely blind. Anatomically, the diseases differ essentially. Macular degeneration begins in the outer layers of the retina, and is limited to the central region. Amaurotic idiocy, on the contrary, begins in the inner layers and extends over the entire retina. By no means, is it especially a macular disease; all regions of the retina are equally affected, but the opacity is most conspicuous about the fovea simply because here the dead ganglion cells are eight or nine layers in thickness as compared with a single layer elsewhere. The strong ophthalmoscopic picture thus produced appears to have engendered an erroneous idea that amaurotic family idiocy is a macular disease. Treacher Collins, who saw F. D. Battin's cases, said the dark spot at the macula was composed of definite pigment granules, and was not due to contrast as in amaurotic idiocy.

**CENTRAL CHORIORETINITIS.** The usual form of central choroiditis bears little resemblance to maculocerebral degeneration. Choroiditis frequently depends upon evident syphilis or tuberculosis. It presents inflammatory phenomena, and the early lesions appear as deeply situated foci of exudation. These are followed by formation of white cicatrices and massing of pigment. The history and course of maculocerebral degeneration have no counterpart in exudative choroiditis.

**UNCLASSIFIED CASES.** Ophthalmic literature contains many unclassified cases, which may or may not belong to the same group as maculocerebral degeneration. The most important are the following by Stock. Although regarded by the author as an independent disease, it is strongly suspected that they were cases of congenital syphilis.

Four children in a childship were affected. No parental consanguinity. After the birth of the first child, which was healthy, and escaped the disease, the father acquired syphilis (statement uncertain), and now is in America. The four patients were normal until they were six years of age, when vision and intellect began to fail. They exhibited posterior iritic adhesions and multiple glandular enlargements. The iritic adhesions disappeared after mercurial inunctions. No paralysis. One case was epileptic. They rapidly became absolute imbeciles, and completely blind. At first the fundus presented no visible change, but later pigmentation of the

retina commenced at the periphery and extended toward the center. The optic nerve and retinal arteries showed no positive evidence of atrophy. Three of the cases died from tuberculosis and the eyes were secured for examination, in one case an hour and a half after death. The findings were of the first importance in their bearing upon all forms of essential retinal atrophy. Optic nerves were normal. Choroid and inner layers of the retina were comparatively free from degeneration. The only certain pathologic change was more or less complete degeneration of the rods and cones.\*

This disease resembled maculocerebral degeneration in that it was aroused to activity during the second dentition and manifested itself by destruction of vision and intellect. It may be differentiated, however, by the presence of other eye lesions, constitutional taint, acute course, absence of optic atrophy, and primary location of the retinal disease at the periphery. The limitation of maculocerebral degeneration to the central zone is alone sufficient to separate the two conditions.

In the ophthalmoscopic picture of maculocerebral degeneration neither the amount of pigment nor its arrangement has much bearing upon diagnosis. The essential point is the presence of "simple" as distinguished from postinflammatory atrophy of the retina limited to the posterior pole.

### *Bibliography.*

- Battin, F. E., Transactions Ophthalmological Society, 1903, XXIII, 386.  
 Battin, R. D., Trans. Ophthalmological Soc. United Kingdom, 1897, XVII, 48.  
 Best, An Hereditary Macular Affection, Zeits. f. Augenh., 1905, XIII, 199.  
 Collins, Treacher, Transactions Ophthalmological Society, 1903, XXIII, 390.  
 Hirschberg, Centralb. f. p. Augenh., January, 1904, p. 12.  
 Knapp, Transactions American Ophthalmological Society, 1870, I, 121.  
 Leber, Unpublished case cited by Stargardt.  
 Mayou, Transactions Ophthalmological Society, 1904, XXIV, 142.  
 Oatman, Arch. of Oph., 1907, XXXVI, 554.  
 Stargardt, Archiv f. Ophthalmologie, 1909, LXXI, 543.  
 Stephenson, Transactions Ophthalmological Society, 1904, XXIV, 144.  
 Stock, Klin. med. f. Augenh., XLVI, 1, p. 225.  
 Vogt, Ueber familiäre amaurotische Idiotie, Monatsschr. f. Psych. u. Neurologie, 1905, Band XVIII, p. 161; Band XXII, p. 495.

**AMAUROTIC FAMILY IDIOCY (Tay's Disease).** Warren Tay, in 1881, was the first to describe a peculiar disease of the cerebrospinal system, occurring exclusively in children and attended by idiocy and blindness. All indubitable cases so far reported have occurred among Hebrews. The

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\*Usually, it is uncertain that microscopic changes found in the rods and cones are due to disease. Post-mortem degeneration and preparing the eye for examination cause these delicate structures to disintegrate.

disease tends to attack several members of a childship at about the same period of life. The child remains healthy until from four to six months of age when it becomes apathetic and sleeps most of the time. Gradually, paresis or paralysis of the general muscular system develops and the patient is unable to sit up or to support the head. Certain muscles may be spastic instead of flaccid. Abnormal sensitiveness to touch and sound (hyperacusis) often appears and when present increases the difficulty of an ophthalmoscopic examination. Occasionally, convulsions occur but are not considered a symptom of the disease. Two cases have been reported in which hearing was lost (Sachs). Coincident with the general nervous affection, eye symptoms develop which are peculiar to amaurotic family idiocy. Both eyes are symmetrically affected. Failure of vision is soon followed by blindness. The pupils are dilated and sluggish and, in the last stage of the disease, irresponsive to light. Rolling the eyes, or nystagmus is common. The ophthalmoscopic picture is characteristic of the affection. The macula is occupied by a broad, whitish disc in the center of which the fovea appears as a reddish spot, producing the same appearance as seen in closure of the central artery. The optic disc becomes pale and ultimately white. It is not surrounded by retinal opacity. In all cases which I have examined the retinal vessels appeared normal. All bodily functions are torpid, especially manifested by pulmonary and gastrointestinal disturbances. With advance of cerebral degeneration the child passes into absolute imbecility. The processes of metabolism are seriously impaired and a condition of extreme marasmus develops which nearly always terminates in death within two years. Sachs has met but one example of the disease which extended beyond two years. In this case other members of the childship were affected. When seen by Sachs the child had attained the age of  $5\frac{1}{2}$  years and death did not appear to be imminent.

ETIOLOGY. The etiology of amaurotic family idiocy is unknown. The most remarkable feature of the disease, as pointed out by Carter, is racial influence. In New York, which has a Hebrew population of a million, I see from one to three cases of the disease yearly, but have never met a suspicious instance among gentiles. This racial peculiarity appears to exclude either toxic, bacterial or accidental origin for the disease. None of the authentic cases has been attributed to syphilis. Consanguinity of parents is noted in some cases and undoubtedly exists in others in which it is denied. It is uncertain whether the disease is congenital or acquired, but probably the former. The suggestion that mother's milk may be

causative is disproved by several of Sach's cases who were fed by strange wet nurses. Sachs considers amaurotic idiocy as some congenital defect which, at a certain age, arrests development. This is a modification of Edinger's *Ersatztheorie*, according to which tissue endurance is too feeble to withstand the physiologic activity of life. A family taint of insanity in both parents has been suggested as causative. Injury of the mother during pregnancy has been reported in several instances of the disease.

**MORBID ANATOMY.** The first anatomical examination was made by Sachs in 1887. There was no evidence of inflammatory action. The brain was unduly firm with the consistence of gutta-percha. The essential histologic feature of the disease was found in the cortex and consisted of degenerative changes in the ganglion cells. Hirsch was the first to show that the degeneration of ganglion cells found in the brain extended throughout the grey matter of the entire central nervous system. Ward Holden in 1898, was the first to demonstrate that the blindness and peculiar appearance in the retina were caused by degeneration of the ganglion cell and nerve fiber layer. It is assumed that the degeneration is primary in the cell and secondary in the fiber or axon. The ganglion cells were increased in size and contained a globular nucleus. Cell membrane and cytotreticulum were intact, but the Nissl bodies had entirely disappeared.\*

The macular opacity seen with the ophthalmoscope has been attributed to edema of the retina, but it is improbable that any real edema exists, the opacity being caused solely by the dead ganglion cells which are eight or ten layers thick in this region, while in other parts of the retina they consist of a single layer. In some cases a fold in the retina involving the macula has been described (Fig. 130), but this is a post-mortem artefact common in the eyes removed many hours after death. Likewise, the presence of a hole in the macula was supposed to explain the red spot, but this also is an artefact produced by rough manipulation or laboratory processes.

I microscopically examined one case of this disease in which the eye was removed eight hours after death.† The macula was thrown into a sharp post-mortem fold (Fig. 130). At the macula the external granular layer was normal; the external reticular layer was not edematous; internal granular and ganglion cell layer were very thick. Owing to the time that

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\*The ganglion cells of the retina undergo rapid post-mortem disintegration, and examinations must be made almost immediately after death.

†From the service of Dr. A. E. Davis, New York Post Graduate Hospital.

elapsed between death and fixation of the eye by preservatives, it cannot be asserted that all the finer cell changes were due to disease.

The optic nerve was atrophic. The atrophy and thinning of the nerve fiber layer being especially marked on the temporal side of the disc (Fig. 131).

**STEREOGRAM 57. AMAUROTIC FAMILY IDIOCY.\*** Right eyeground of male infant, aged 10 months; firstborn; Russian Hebrew; parents are second cousins; the child is apathetic and appears to be blind; pupils are dilated and very sluggish; a paretic condition of the muscular system is shown by inability of the child to sit up or to support the head, which droops. *Ophthalmoscopic Picture.* The fovea appears as a maroon colored spot, surrounded by a wide zone of greyish-white retina. The optic nerve is pallid, especially on the temporal side. The retinal vessels appear normal and a number of small vessels around the macula, ordinarily invisible, are brought into view by contrast with the white retina.

**DIAGNOSIS.** Amaurotic family idiocy is diagnosticated by the presence of characteristic fundus changes in a paralytic infant, and by the history. It is said that the cerebrospinal degeneration may precede the eye symptoms which, however, develop later. The ophthalmoscopic picture differs from that of central artery closure in that the vessels are not narrowed, or the retina opaque around the nerve.

### *Bibliography.*

- Tay, T. Oph. Soc., vol. I, 1881, 55.  
 Sachs, Jour. Ment. & Nerv. Dis. 1887, 541; 1892, 1903. N. Y. Med. Jour. 1896. Deutsch. Med. Woch. 1898; 1903. Posey and Spiller, "The Eye and Nervous Dis." p. 532.  
 Carter, A. of Oph. XXIII, 1894.  
 Edinger, Parsons, Path. of the Eye, IV, 1368.  
 Hirsch, Jour. of Ment. and Nerv. Dis., 1899, 538.  
 Hirsch and Holden, Jour. Ment. and Nerv. Dis. 1898, XXV.  
 Holden, Trans. Am. Oph. Soc., 1898, 405.  
 Davis and Oatman, Contrib. to Med. & Surg. 25th Anniver. N. Y. Post Grad. Med. School and Hospital, 1908.

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\* From the clinic of Dr. P. C. Jameson, Brooklyn Eye and Ear Hospital.

## CHAPTER X.

### MISCELLANEOUS AFFECTIONS OF THE RETINA.

CYANOSIS RETINÆ. Marked distension and tortuosity of the retinal vessels unattended by hemorrhage, exudation or edema, occurs in diseases which produce general venous stasis. This retinal condition is most frequently seen in connection with the general cyanosis incident to an open foramen ovale in which there is admixture of venous with arterial blood within the heart, and also in cases of pulmonary stenosis. Congenital cyanosis has been classed as a disease under the name of *Morbus Cæruleus*, or blue disease, and the subjects of the affection are known as blue children. The congestion seen in the retina exists in other organs; thus, enlargement of the liver and spleen, atelectasis, pleural effusion and edema of the brain are frequent. The disease is recognized by the peculiar livid hue of the skin, which varies from a dusky tinge to a deep purple color. Although the entire surface of the body is affected, the discoloration is most marked on the lips, cheeks and eyelids. The skin is dry and the extremities cold. Palpitation and dyspnea are always present and are greatly increased by excitement or exercise. Somnolence is a common symptom. Polycythemia develops in cases of pulmonic obstruction. Increase in the number of red blood corpuscles is proportionate to the cyanosis, the average number present being about 7,000,000 (Tyson). A corresponding increase occurs in the amount of hemoglobin. The leucocytes are not increased in numbers.

The duration of life varies. One half of all cases die in the first year. Other cases survive until adult life. Litten mentions a case which lived to the age of 25 years. There is, of course, no chance of recovery. The most dangerous periods of life for cyanotic cases are the first respiratory act, the period of weaning, first dentition and during any intercurrent disease. In the later period of the disease the retinal vessels may become diseased and hemorrhages occur. Posey reports a case presenting the picture of a neuroretinitis.

Congestion of the retinal vessels producing the ophthalmoscopic picture of "cyanosis retinæ" has been observed in emphysema by Stephen Mackenzie. In one of Hirschberg's cases of general and retinal cyanosis,

there were pulmonary emphysema and hypertrophy of the heart which were due to kyphoskoliosis. The condition of intense retinal hyperemia has been seen during the height of an epileptic seizure and also in certain intoxications (Stergm. 60).

STEREOGRAM 58. RETINA IN CONGENITAL CYANOSIS (Cyanosis Retinæ). Right eyeground of a "blue boy," eleven years of age. Since birth the skin has been cyanotic. The blue color is especially marked on the lips and eyelids. Owing to dyspnea he has never been able to play like other children. A physical examination indicated patent foramen ovale and pulmonic stenosis. Refraction is emmetropic. Vision 20/20.

OPHTHALMOSCOPIC EXAMINATION. The entire fundus has a deep red color. The optic disc appears reddened except in the physiologic excavation. Both veins and arteries are very tortuous and somewhat dilated. The arteries are not sclerosed, as shown where they cross over a vein without indenting the latter.

DIAGNOSIS. The diagnosis of cyanosis retinæ rests upon the presence of general cyanosis. The appearance of the fundus is not very characteristic of the disease. For example, were this eye hyperopic it could not be ophthalmoscopically diagnosticated from the picture sometimes presented by that condition. Arteriosclerosis may be excluded by the perfectly normal appearance of the vessels. Cyanosis is differentiated from inflammatory conditions by normal vision and absence of hemorrhage, exudation and retinal edema.

### *Bibliography.*

- Von Ammon, Nor. & Oliv., vol. III, p. 433.  
Stephen Mackenzie, Trans. Oph. Soc., 1882-3.  
Posey, Trans. Am. Oph. Soc., X, p. 632.  
Hirschberg, Arch. f. Oph. LIX, p. 131.  
Litten, Deutsch. Med. Wochenshrf., 1887, p. 144.  
Tyson, Trans. Am. Oph. Soc. 1908.

TRAUMATIC EDEMA OF THE RETINA (Commotio Retinæ). Temporary blindness varying in duration from a few moments to several hours nearly always follows violent concussion of the eye. This condition of traumatic amblyopia may appear and disappear without visible alteration in the retina. In other cases deterioration of vision persists and the retina undergoes characteristic changes consisting of an opacity which extends more or less completely over the posterior polar zone. In well marked cases the appearance of the retina resembles somewhat the condition which follows closure of the central artery. The opacity, which is



due to edema of the retina, is most conspicuous where the membrane is thickest, namely, around the fovea centralis and optic disc. The fovea itself, however, is the thinnest portion of the retina and may permit the choroid to shine through and produce a "cherry-red" spot. In other cases the edematous retina may overlie the fovea so that it is nearly or completely obscured. Sometimes the retina at the fovea is torn and an actual hole exists. This accident usually occurs in myopic eyes. In all cases of commotio retinæ which I have observed the retinal opacity at the macula was more sharply outlined than in closure of the central artery. The blood vessels usually are dilated and the perivascular lymph channels may be distended. The visible retinal changes vary from an almost imperceptible cloudiness at the macula to dense opacity and, in rare cases, hemorrhages.

The term "commotio retinæ" formerly was applied to any obscure case of visual impairment following injury to the head. Further investigation established edema of the retina, especially of the nerve fiber layer, as the chief anatomic alteration, although the mechanism by which the edema is produced is not fully determined. Berlin, who first separated these cases, thought the retinal edema was caused by suprachoroidal hemorrhage, a view soon abandoned. The majority of writers now agree that the first result that follows a blow to the eye is retinal ischemia due to spasm of the blood vessels. The angiospasm is immediately followed by vascular dilation and transudation of serum. Denig, as the result of animal experimentation, states that a violent blow drives the vitreous against the retina, rupturing the internal limiting membrane and forcing the vitreous fluid between the nerve fibers and along the fibers of Müller. Traumatic retinal edema may occur with rupture of the choroid or other effects of blunt force. The most commonly associated conditions are opacity of the cornea, miosis, mydriasis, lacerations of the iris, dislocation or opacity of the lens and hemorrhage.

In the following case the macular changes found on microscopic examination were regarded as those of traumatic edema of the retina.

A man received a nonperforating blow in the eye. Twenty-four hours later the eye was glaucomatous with widely dilated pupil. The lens was dislocated forwards. Vision reduced to movements of the hand. Details in the fundus were indistinguishable although the reflex from the macular region was much lighter in color than from other regions. Iridectomy and extraction failed to relieve the condition and the eye was enucleated fourteen days after the injury. *Microscopic examination.* The iris is thrown into folds and lies in the angle of filtration where it is



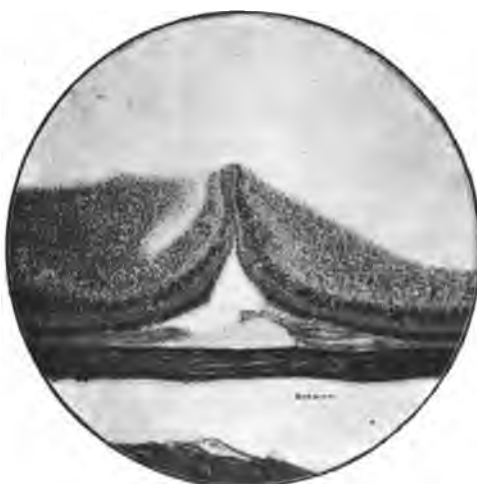


Fig. 130.

Fig. 130—Amaurotic Family Idiocy. Macular Opacity Due to Death of the Thick Layer of Ganglionic Nerve Cells. The Retinal Fold is an Artefact.

Fig. 131—Amaurotic Family Idiocy. Optic Atrophy, which is Most Marked on the temporal Side of the Disc, the Left Side in the figure.



Fig. 131.



Fig. 132.

Fig. 132—Glaucoma Following Injury. The Iris, Thrown into Folds and in Contact with the Cornea, Occludes the Filtration Angle.

closely pressed against the cornea (Fig. 132). The condition of the macula is shown in Fig. 20. The retina is edematous, the edema being limited to the nerve fiber and internuclear layers. The fovea is widened and contains a serous effusion.

In commotio retinæ it is probable that minute changes occur in the nervous tissue not demonstrable with the microscope. Central vision is always impaired and the visual fields are somewhat concentrically contracted.

In cases where the traumatism results only in edema the prognosis is good and vision is fully restored. Within a week or two the retinal opacity disappears, at which time white spots situated in the deeper layers of the retina come into view. In a short time these also are removed and no trace of the effusion remains. Exceptionally, slight pigmentation results, in which case it is supposed that the choroid participates in the disturbance. When concussion results in actual rupture of the retina and a hole forms at the macula, the prognosis is less favorable. The hole does not always result directly from concussion, but may develop later from excessive edema. Holden reports a case in which three weeks after a blow the retina was intact, but two months later a typical hole developed at the macula.

STEREOGRAM 59. TRAUMATIC EDEMA OF THE RETINA (Commotio Retinæ). Right fundus oculi of a barkeeper, 33 years of age. Seven days ago he received a violent fist-blow on the right temple, immediately followed by loss of vision in the right eye. At the present time, with this eye he can count fingers at five feet. Retinal edema is marked around the nerve and macula and slightly developed throughout the rest of the posterior zone. The macula is occupied by a sharply defined, rounded, grey disc in the center of which the fovea appears as a dark red spot. Around the nerve the edema is greatest on the nasal side where it nearly hides the outline of the disc. The veins are full and darkened. Arteries and veins on and near the disc are bordered by delicate white lines, supposed to represent an effusion into the perivascular sheaths. The nerve and vessels present the appearance of commencing neuritis.

Improvement in vision immediately ensued. The edema rapidly disappeared, the macular spot grew indistinct and within a week the fundus exhibited little evidence of injury.

DIAGNOSIS. In the great majority of cases a history of injury at once leads to the correct diagnosis. Occasionally, however, the true history is suppressed. In both traumatic edema and closure of the central artery

there may be retinal opacity and a red spot at the macula. In arterial closure the arteries are greatly narrowed or invisible while in traumatic edema the vessels are overfull. In traumatic edema the macular opacity is apt to be more sharply defined than in closure. Amaurotic idiots present macular opacity and a red fovea, but the subjects of this disease always are infants affected by a general paresis.

### *Bibliography.*

- Berlin, Kl. F. f. Augk., XI, 1873: Graefs Saemisch, VI, 1880.  
 Ward Holden, Trans. Oph. Sec. N. Y. Acad. Arch. of Oph. XXXIII, 1904, p. 190.  
 Denig, Arch. of Oph. XXVI, 1897, p. 377.

### TOXIC AMAUROSIS AND AMBLYOPIA.

**STEREOGRAM 60. RETINITIS IN WOOD ALCOHOL POISONING.** (Congestive Edema of the Retina).\* Left eyeground of a young man blind from acute, methyl alcohol poisoning. After working three weeks shel-lacing the interior of beer vats, where he inhaled the concentrated fumes of wood alcohol in which the shellac was dissolved, he was seized with violent vomiting and 24 hours later became totally blind. The pupils were widely dilated and unresponsive to light. About one month later he could count fingers with the left eye but perception of light never returned in the right. Both optic nerves gradually atrophied and permanent blindness ensued.

The ophthalmoscopic picture, taken in the early stage, indicates an inflammatory edema of the retina. Both arteries and veins are dilated and tortuous. Extensive effusion of serum into the retina has occurred along the larger vessels. There is no optic neuritis, although edema from the retina has extended over the upper and lower margins of the disc. Were this nerve swollen, the arteries would be compressed and narrowed.

**DIAGNOSIS.** The ophthalmoscopic picture is not characteristic and the diagnosis in wood alcohol poisoning is made chiefly from the history. In acute wood alcohol amaurosis, blindness develops rapidly in both eyes, hours or days after a debauch. Pupils dilated and fixed. The fundus picture varies, there may be, (1) normal nerve and retina; (2) congested nerve and retina; (3) retinal ischemia. Usually there is temporary improvement followed by blindness or constricted fields. In uremic amaurosis the pupils usually respond freely to light while in wood alcohol poisoning they are immobile. If uremic coma coexists with amaurosis, casts and

\* From the service of Dr. David Webster, Manhattan Eye, Ear and Throat Hospital.



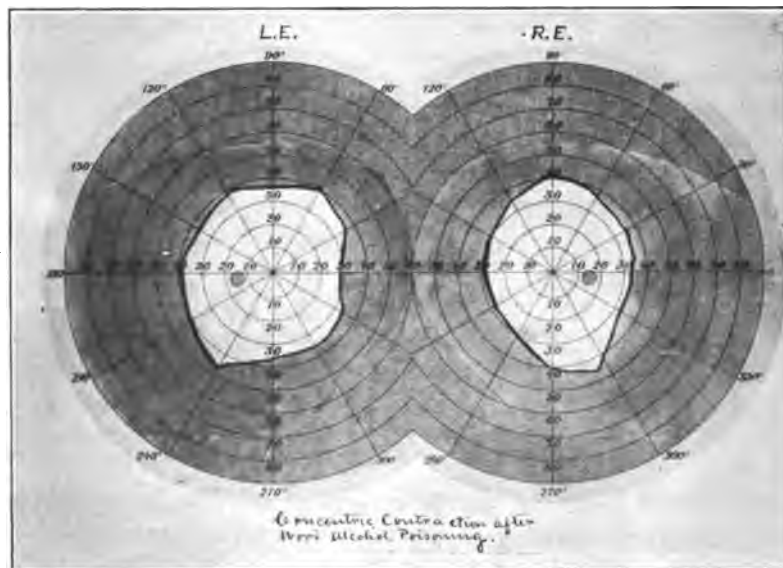


Fig. 133 (a).

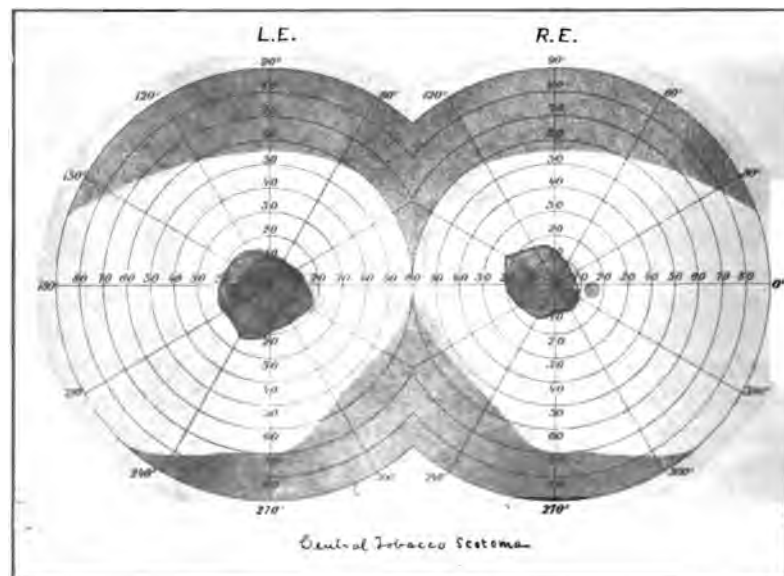


Fig. 133 (b).

albumin may temporarily disappear from the urine. Retinitis may or may not coexist. Amaurosis from cerebral or spinal disease develops gradually and is steadily progressive.

Acute poisoning by wood alcohol is not rare. Usually, it follows a debauch upon some cheap liquor composed largely of methyl alcohol, or from prolonged inhalation of its fumes. In the lesser degrees of poisoning central scotoma may be demonstrated. In late cases concentric contraction of the white fields may exist. Blindness may not develop for several days after ingestion of the poison. Paralysis of the face or limbs and loss of intelligence are not uncommon in wood alcohol poisoning. Temporary improvement of vision often ensues, followed by total blindness. Other cases recover with more or less contraction of the fields (Fig. 133a).

Tyson\* has called attention to chronic toxic amblyopia which is not uncommon in those who work with colors or varnishes dissolved in wood alcohol.

As demonstrated by Holden, the blindness produced by methyl alcohol is due to destruction of the ganglion cells in the retina. In addition to its action on nerve tissues, it exerts a powerful influence on the retinal blood vessels manifested, however, by widely divergent ophthalmoscopic pictures. In one type a condition of retinal anemia with narrowed vessels exists, in another the vessels are dilated and the retina acutely congested. Again, there may be absolute blindness without visible fundus changes.

**CHRONIC ALCOHOLISM.** Amblyopia due solely to chronic alcoholism is extremely rare. It differs in no respect from tobacco amblyopia, except that other symptoms of alcoholism may coexist.

**TOBACCO AMBLYOPIA.** The intemperate use of tobacco and alcohol usually go hand in hand but, clinically, this form of amblyopia occurs almost exclusively among those who for years have been excessive consumers of tobacco. The symptoms develop slowly. Both eyes are uniformly affected. The first complaint is of foggy vision and of "dazzling" when in a strong light. Day blindness (nyctalopia) is a prominent symptom and its presence always suggests tobacco poisoning. Gradually, central vision deteriorates and after a time reading becomes impossible although the peripheral fields of vision are retained. The visual fields present a central color scotoma for red and green with a relative central scotoma for white (Fig. 133b). The scotoma in tobacco amblyopia corresponds to the distribution of the maculopapillary bundle of nerve fibers, *i. e.*, a horizontally oval area which just embraces the macula and optic

\*Tyson, Arch. of Ophthalmology, Sept. 1912.



nerve. If the patient persists in the use of tobacco the relative white scotoma becomes absolute and central vision is lost. The peripheral fields, however, are not affected and complete blindness does not occur. If the use of tobacco is discontinued when toxic symptoms first develop, recovery may be expected.

The pathologic lesion is generally regarded as a primary inflammation of the maculopapillary bundle of nerve fibers in the optic nerve. This cannot be considered as fully established, particularly as blindness in methyl alcohol poisoning is supposed to be caused by destruction of the ganglion cells in the retina.

**IODIFORM POISONING.** Poisoning by iodiform, which at one time was common, produces eye symptoms almost identical with those of alcohol and tobacco.

**QUININE AMBLYOPIA.** Quinine in poisonous doses produces extreme ischemia of the retina and blindness. As a rule, vision returns, although if poisoning has been severe permanent, concentric constriction of the visual fields results. In experimental poisoning with quinine, Holden found narrowing of the retinal vessels and degeneration of the ganglion cells. It is not clear whether the ganglion cell changes were due to direct action of quinine or to malnutrition from anemia of the retina. Parsons concluded from experiments on animals that the retinal ischemia in quinine poisoning was due to withdrawal of blood from the surface and its accumulation in the great abdominal veins.

**SALICYLIC ACID** may produce temporary amblyopia resembling quinine poisoning as described above. Other drugs which may produce amblyopia with a pale disc and *retinal anemia* are felix mas, oil of wintergreen, cocaine, ergot, antifebrine, acetanalid, adrenaline, nitrobenzol and chlorate of potash.

Amblyopia, usually accompanied by *congestion* of the nerve and retina, occurs in poisoning by phosphorus, scamonium, pomegranate, colocynt, ptomaines and aniline oil. The last named may exhibit a violet fundus and black vessels.

Amblyopia has been reported from overdoses of sulphuretted hydrogen, carbolic acid and other drugs. The intramuscular injection of arsenic and anilin preparations for cure of syphilis has been followed by optic atrophy.

The fundus picture of *lead amblyopia* is inconstant and varies from choked disc with retinal hemorrhage to blindness without ophthalmoscopically visible changes.





Fig. 134.

Fig. 134—Retinitis Proliferans. Dense Masses of Connective Tissue in the Inner Retinal Layers.

Fig. 135—Retinitis Proliferans. Thin Membrane of Connective Tissue arising from the Retina and Extending Forward into the Vitreous.



Fig. 135.

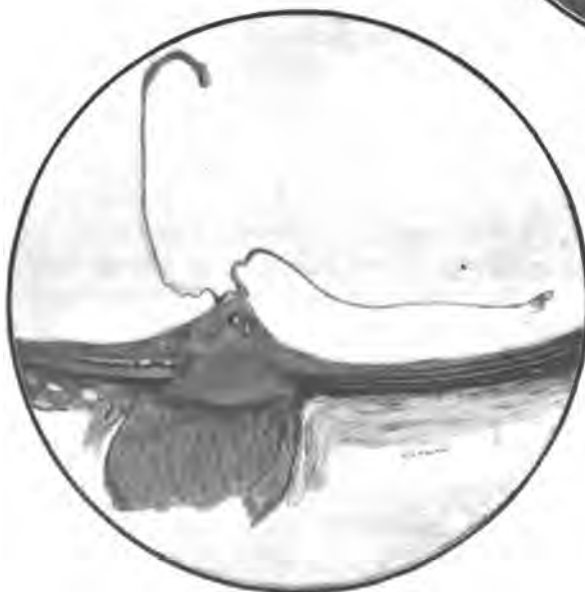


Fig. 136.

Fig. 136—Retinitis Proliferans. Connective Tissue Formation on Disc. Thick Membranes Originating therefrom are Continued Forward into the Vitreous.

Colored vision (*chromotopsia*) may be caused by drugs. Santonin usually produces yellow vision, less frequently, red or green vision. Chromotopsia may be caused also by chromic acid, picric acid, amyl nitrite, cannabis indica, digitalis, carbonic acid, tobacco and coffee. Temporary scotoma may be due also to coffee.

**RETINITIS PROLIFERANS.** In 1869 Jager illustrated, and in 1876 Manz named, the condition known as retinitis proliferans. The essential feature of this disease is the presence of dense masses of connective tissue (Fig. 134) or of thin membranes, which originate in the retina and extend forward into the vitreous (Fig. 135). These sheets of connective tissue usually arise from the disc or its vicinity and are attached anteriorly to the ora serrata. Nearly all authors regard hemorrhage as an essential preliminary in the formation of retinal membranes, which are supposed to be metamorphosed fibrinous coagula. As stated by Flemming, however, some additional factor is required to convert blood clots into membranes. Proliferation of neuroglia, especially of Müller's fibers, has been accepted by some as the source of the new tissue, but extensive proliferation of neuroglia is an improbable event. A more reasonable origin for the membranes is found in proliferation of the connective tissue (*adventitia*) which surrounds the vessels on the disc (Wehrli, Römer, Parsons). Under proper stimulation this tissue, like connective tissue elsewhere, actively proliferates. In a similar manner, connective tissue around a retinal vessel may produce enormous hypertrophy of the inner layer of the retina (Fig. 134).

It is not unusual to find fibrous membranes in eyes which have been enucleated for injury attended by intraocular hemorrhage, particularly when a foreign body is retained within the eye. Figure 136 represents a section from an eye in which a piece of stone was imbedded for eight months in the posterior extremity of the ciliary body. There is enormous proliferation of connective tissue on the disc. The same tissue is continued into the vitreous in the form of thick membranes. In this case the foreign body which stimulated proliferation was remote from the nerve.

Flemming's idea that the hyaloid membranes are concerned in retinitis proliferans appears true for certain cases in which the vitreous septa appear to form a foundation on which extraneous matter is deposited (Fig. 137).

Figure 137 is from a case of typical traumatic retinitis proliferans preceded by hemorrhage.\* A piece of steel entered through the cornea and lodged in the retina near the disc. The intraocular hemorrhage which

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\*From a case which I microscopically examined for Dr. Dunbar Roy, of Augusta, Ga.

resulted prevented an ophthalmoscopic examination. The steel was removed by magnet and the wound healed. Six months later the eye was enucleated for a burn of the cornea. The microscopic examination showed a funnel-shaped fibrous membrane, the apex of which was attached to the retinal wound, near the nerve, and the expanded edge to the ora serrata (Fig. 138). It is interesting to note that a band of connective tissue extended between the disc and posterior attachment of the membrane.

Retinitis proliferans is uncommon. Weeks, among 24,000 patients met but two examples. The condition may be due to traumatism or to some constitutional dyscrasia. Weeks speaks of syphilis as a common cause. Spontaneous cases occur in ill-nourished young people, usually males. Frequently, both eyes are affected. If spontaneous cases are preceded by hemorrhage some form of vascular degeneration may be assumed. Exceptionally, contraction of the membranes leads to detachment of the retina (Scott).

The prognosis is unfavorable. Usually the eye becomes soft and phthisical although, as in other forms of intraocular hemorrhage, glaucoma may develop.

STEREOGRAM 61. RETINITIS PROLIFERANS. Right eyeground of a man aged 28 years. He is a poorly nourished specimen of a city degenerate; blood pressure 178 mm. Hg.; surface arteries palpable; urine contains a trace of albumin. Has often been severely clubbed by the police and beaten in fistic encounters. Following such an encounter two years ago, vision became poor in the right eye. *Ophthalmoscopic Examination.* The left eye contains no membranes but a small hemorrhage has occurred between the macula and disc. The right eye contains a number of thin membranous formations, attached to the disc, some of which extend forward beyond the limits of the ophthalmoscopic field. The upper of these new formations appears to be a solid mass of connective tissue covered with adventitious blood vessels. The membranes are greyish-white in color, very dense at the disc but thin and semitransparent anteriorly. They are immovable and do not float about in the vitreous when the eye is moved.

DIAGNOSIS. The ophthalmoscopic picture of retinitis proliferans is unmistakable. These membranes are distinguished from detachment of the retina by their whitish color, membranous form and attachment to the nerve. Occasionally, retinitis proliferans leads to detachment. These formations differ from vitreous opacities of inflammatory origin by their color and immobility.



Fig. 137.

**Fig. 137—Retinitis Proliferans.** The Fibrous Septa of the Vitreous appear to have formed a Foundation on which Extraneous Matter has been Deposited.



Fig. 138.

**Fig. 138—Retinitis Proliferans.** From same specimen as Fig. 137. The Membranous Formation, arising from near the Nerve Head, expands Funnel-shaped to the Ora Serrata and might be mistaken for a Visible Hyaloid Canal.



*Bibliography.*

- Jäger, Handatlas, 1869.  
 Manz, A. f. Oph., XXII, 3, 1876; XXVI, 2, 1880.  
 Flemming T. O. Soc., XVIII, 1898.  
 Wehrli, A. f. Aughn., XXXVII, 1898.  
 Römer, A. f. Oph., LII, 3, 1901.  
 Parsons, Path. of the Eye, vol. 2, p. 614.  
 Weeks, Text Book, 1910, p. 458; Trans. Am. Oph. Soc. 1897.  
 Scott, T. O. Soc., XXIII, 1903, p. 71.

RETINITIS STRIATA AND ANGIOID STREAKS IN THE RETINA. All forms of striate conditions in the fundus have been described as retinitis striata but this name should be reserved for two fairly well defined clinical pictures, now designated respectively as retinitis striata and angioid streaks.

Retinitis striata is the term applied by Nagle, in 1864, to a peculiar affection apparently allied to retinitis proliferans. It is characterized by the presence of branching striæ in the retina situated behind the retinal vessels and in front of the tessellated epithelium. The streaks vary in width from one to three times the diameter of a primary retinal vessel, are bluish- or yellowish-white in color and not infrequently bordered or covered by a dark brown pigment. The course pursued by the striæ is inconstant; they may radiate from the disc in the most eccentric directions (Dunn) or they may traverse the fundus diagonally in long, straight lines, the general direction, however, being from periphery to center. The vessels may appear normal or sclerosed; in some cases they have been reported as extremely tortuous (Gloor; Schilling). In most reports the vitreous is described as clear, occasionally as turbid. The disease does not appear to have any constant subjective symptoms such as disturbance of the color sense, hemeralopia, nyctalopia, etc. Usually, vision is considerably lowered and the visual field irregularly contracted, but these symptoms appear to bear a relation to the amount of mechanical destruction of tissue. The retina may or may not exhibit opacity. This variety of striate retinitis usually is confined to one eye and in many cases a traumatic origin can be traced.

Angioid streaks in the retina is a condition closely related to, if not identical with, the retinitis striata of Nagle. The first case was reported by Plange, to which Herman Knapp immediately added another and named the disease "angioid streaks in the retina." In this type of the affection the streaks occupy the same position beneath the retinal vessels and pursue a similar course as in retinitis striata. They differ, however, in that they are narrower and consist of brownish pigment instead of white tissue. An ex-



ception as regards the color of the streaks existed in a case by Stephenson in which the brown stripes were bordered on one side by a broader white stripe, suggesting a transformation of angioid streaks into white striæ (retinitis striata). Angioid streaks are more frequently bilateral than retinitis striata. Both types belong to early adult life, while some cases appear to be congenital.

The identity of retinitis striata and angioid streaks remains to be established, but it is at least probable that both conditions originate from blood extravasated into the retina. That this is the origin of angioid streaks has been established by both Holden and deSchweinitz, who have watched the transformation of blood into brown striæ. Holden assumes that the affection called retinitis striata like that called angioid streaks, arises through periphic hemorrhages diffused in a linear manner through the deep layers of the retina where they undergo various sorts of metamorphosis. Inasmuch as the retinal epithelium is not disturbed, the only other apparent source for the brown pigment is blood. Plange ascribed the striæ to hemorrhages which excited secondary hyperplasia of Müller's fibers. Dunn suggested an inflammatory origin but observing no change in the streaks after prolonged observation, he accepted a hemorrhagic origin for the condition. Walser thought that retinal striæ might be due to colloid excrescences, intrauterine hemorrhage or, what he regarded as most probable, to folds in the retina. Pretori believes in the congenital origin of the affection. Striæ may form in cured detachments of the retina, but reattachment is a much more infrequent cause for retinal striation than is generally supposed. While there is little doubt of the hemorrhagic origin of retinitis striata, it is difficult to understand why blood should assume a linear form in the retina. True, Schwalbe and His have described retinal lymph spaces radial with the disc, but the existence of such channels is doubtful. It is highly probable that some cases of retinal striæ may originate in natal hemorrhage occurring while the infant's thorax is compressed in the parturient canal (compression, or traumatic asphyxia).

STEREOGRAM 62. RETINITIS STRIATA. Left eyeground of a woman aged 23 years. Vision in the right eye is 20/20 and in the left eye 20/50; states that vision in the left eye has always been defective.

OPHTHALMOSCOPIC PICTURE: Media clear; optic nerve and retinal vessels appear normal; the distinguishing feature in the fundus is a number of white and brownish striæ, situated in the retina behind the vessels but in no way connected with them. The striæ are white, although in many places they are overlaid or bordered by brown pigment. Two stripes start from

the edge of the disc, one of which proceeds upward and the other downward. At first they are straight and brown, but soon branch and exhibit an underlying white structure. These striæ are joined by others of a yellowish-white color and free from pigment, which extend diagonally across the retina.

The origin of the streaks in this retina can only be surmised, but I am disposed to regard them as the residue of hemorrhages which occurred either during birth or in early infancy.

**DIAGNOSIS.** In retinitis proliferans the new-formed tissues are situated in front of the retinal vessels, while in retinitis striata they are behind them. Retinal streaks due to reattachment of a detached retina are diagnosed only by an undoubted history of preceding detachment as in Görlitz's case. From a preconceived but mistaken view of their etiology, many cases of retinitis striata have been wrongly attributed to detachment. A very narrow, branching rupture of the choroid might be mistaken for retinitis striata, but may be distinguished by its crescentic form and by its situation which is concentric to the outline of the disc.

### *Bibliography.*

- Nagle, *Klin. mon. f. Aughn.*, 1864, p. 394.  
 Schilling, A. f. *Aughn.*, XLIII, 1901. *Arch. of Oph.* XXXII, 1903, 219.  
 Gloor, A. f. *Aughn.* XXXV, 1897, 328. *Klin. m. f. Aughn.*, XXXVI, 1898, 137.  
 Plange, A. f. *Aughn.*, XXIII, 1891, 78.  
 Herman Knapp, A. of *Oph.*, XXI, 289.  
 deSchweinitz, *Rep. Am. Oph. Soc.*, 1896; A. of *Oph.*, XXV, 1896, p. 439.  
 Görlitz, *Kl. m. f. Aughn.*, XXXV, 1897, 361.  
 Dunn, A. f. *Aughn.*, XXXIV, 1897, 294.  
 Stephenson, *Trans. Oph. Soc.*, XII, 1892, 140.  
 Holden, *Arch. of Oph.*, XXIV, 1895, 278.  
 Walser, A. f. *Aughn.*, XXXI, 1895, 345.  
 Pretori, *Beitrag. f. A.* XXIV., 3, 1898, 393.  
 Schwalbe, *Lehrbuch d. Anat., d. Auges*, 1887, 99-111.

**METASTATIC RETINITIS (Endogenous Ophthalmitis).** Infection of the retina is caused either by exogenous bacteria introduced through perforations of the globe or by endogenous material carried to the eye through the blood channels. Exogenous retinitis does not come within the scope of this work. Metastatic ophthalmitis may develop in the course of puerperal fever, surgical sepsis or other microbic diseases. All parts of the eye may be attacked, but in this discussion only retinal metastases will be considered (see metastatic choroiditis). Two principal forms of metastatic retinitis are recognized: (1) A progressive, inflammatory process

which terminates either in purulent panophthalmitis, phthisis bulbi or inflammatory detachment of the retina; (2) Septic retinitis of Roth, distinguished by white spots and hemorrhages into the retina and by its non-progressive character.

The progressive, inflammatory type of metastatic retinitis commences in the retina, although the ciliary body is always involved. The condition develops rapidly. White spots and hemorrhages appear in the retina, the vitreous becomes turbid, total detachment of the retina quickly develops and within twenty-four hours vision is abolished. The attack may be accompanied by pain. From this point one or two courses may be pursued, namely, suppuration or resolution. Suppuration is indicated by edema of the lids and purulent iridocyclitis culminating in bulbar abscess. In certain cases the contents of the eye break down and form pus. Spontaneous perforation, however, may not occur, and the globe may remain in the condition of a "cold abscess," subject to inflammatory exacerbations. In cases which terminate by resolution, the detached retina, agglutinated by inflammatory exudates, forms a yellowish-white, immobile mass behind the lens. Proliferative processes may develop in the retina and new blood vessels form on its surface, in which event it closely resembles a malignant neoplasm and hence is known as "pseudoglioma" (Stergm. 77). In children this condition requires to be differentiated from true glioma (page 286). The intraocular tension is diminished in proportion to the destruction wrought to the ciliary processes. In many cases the tension remains normal. If iritic adhesions exist sufficient to obstruct the outflow of aqueous, secondary glaucoma develops.

**PATHOLOGY.** Metastatic retinitis probably is always caused by bacterial emboli in the capillaries, the results varying according to the virulence of the micro-organism or degree of immunity conferred upon the tissues. Failure to demonstrate bacteria in the tissues is not evidence against the bacterial origin of the process, inasmuch as these organisms may disappear in the last stages. Treacher Collins regards the retina as the usual starting place of metastatic endophthalmitis, but difficulty is experienced in finding an example limited to the retina.

In the following case the uvea did not appear to be involved in the active inflammatory process.\*

Right eye of an infant 9 months of age. Measles prevailed in the family during the period of the mother's gestation and parturition. The

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\*From the service of Dr. A. E. Davis, N. Y. Post Graduate Hospital. The eye was sent to me for examination.

mother thinks the child was born blind. A yellowish-white mass was seen behind the lens of each eye. The right eye was enucleated. Microscopic examination showed that the intraocular mass consisted of the totally detached retina which was connected by plastic adhesions to the ciliary body and lens. Aside from this the ciliary body contained no evidence of inflammatory action (Fig. 117).

Frequent opportunities occur to study the early stages of acute suppurative retinitis developing after exogenous infection from perforating wounds and corneal ulcers. The infection usually is implanted in the ciliary body. Leucocytes appear in the retina first at the ora serrata and at about the same time around the optic disc, traveling from the anterior part of the eye backward along the surface of the retina and also through the hyaloid canal. They soon infiltrate the retina and ensheath the vessels, particularly the veins. There is intense inflammatory edema of the retina which is quickly detached by the accumulated fluid. The effusion is rich in proteids, which are precipitated by reagents employed to harden the eye and appear under the microscope either as granular deposits, fibrinous coagula or rounded hyaloid masses. If suppuration does not ensue, these exudates may pass through the degenerative processes which terminate in fatty or calcareous deposits. From the retina the purulent process passes to the vitreous and choroid. The contents of the eyeball may become necrotic and be discharged as pus or undergo organization and atrophy. The metastatic retinitis of meningitis and measles tends to terminate in organization rather than necrosis. If the retina undergoes proliferative inflammation, collections of lymphoid and giant cells may be found. Axenfeld states that 1.3 per cent of the cases which do not suppurate excite sympathetic inflammation. This statement is important and should be either confirmed or disproved.

**SEPTIC RETINITIS OF ROTH.** Roth first reported that in septic conditions a retinitis occurred which bore some resemblance to that observed in albuminuria. In this condition the exterior of the eye usually appears normal. Exceptionally, there are hemorrhages into the conjunctiva and slight chemosis. Pupillary reactions are normal; the media are clear; the nerve head is neither reddened nor swollen; round and oval white spots appear in the posterior zone, distributed about the disc. These spots rarely appear in or near the macula or anterior to the equator, nor do they follow the vessels but when in contact with the latter the spot lies in front. They manifest no tendency to enlarge or to coalesce. Aside from the spots there is no opacity of the retina. Superficial hemorrhages, dis-

tributed in a manner similar to the spots, but not connected with them, usually are present. Sometimes the hemorrhages contain white centers, and Lenhartz has observed these in perfectly fresh extravasations. White spots may occur without hemorrhage or hemorrhages without white spots, either being sufficient to establish a diagnosis of septic retinitis. Litten thought that retinal hemorrhage was the immediate precursor of death but Wilbrand and Saenger have observed it weeks before. Metastatic ophthalmitis of all types occurs in one eye about twice as frequently as in both.

**PATHOLOGY.** The propriety of considering Roth's septic retinitis as only a variation of the severe type of metastatic retinitis is not generally conceded. That it is caused by toxins generated in remote parts of the body remains to be proven. In the meantime, micro-organisms have been demonstrated in the lesions and retinal blood vessels of typical cases. Axenfeld-Goh report a fatal case of ulcerative endocarditis with septic retinitis. Masses of pneumococci were found in the recent retinal deposits, but in the older spots the cocci were degenerating or had disappeared. One small retinal vein was filled with masses of bacteria. In the retina these bacteria possess but slight virulence. Wilbrand and Saenger describe a case in which the retinal vessels were filled with streptococci. Grunert reports a case presenting both types of metastatic retinitis. In the right eye was a severe iridocyclitis with vitreous abscess, and in the left eye a typical septic retinitis of Roth. In the latter case the vitreous remained clear and the appearance of the spots was unchanged at the time of death. Reports upon the histologic changes found in Roth's spots indicate their localized character. Slight edema and loss of nuclear staining may exist in the adjacent retina. In fresh cases Grunert found the spots to consist of sharply defined accumulations of round cells. Wilbrand and Saenger found them to be composed of massed leucocytes. Nearly all reports mention the presence of so-called "ganglionic nerve fibers," which may be regarded as only altered leucocytes.

**DIAGNOSIS.** The ophthalmoscopic diagnosis between Roth's spots and miliary tubercles in the choroid is difficult. Tubercles have a reddish or yellow color, while the septic nodules are white. Retinal hemorrhages are more common in sepsis than in miliary tuberculosis. An optic neuritis would suggest cerebral tubercle. The solitary choroidal tubercle is much larger than Roth's spots.

*Bibliography.*

- Axenfeld, A. f. O. Oph. XL, 4, 1894.  
Goh. A. f. O. XLIII, 1, 1897.  
Grunert, Bericht d. Heid. Oph. Gesl. 1903, 338.  
Lenhartz, Sp. Path. u. Therp. v. Nothnagle, Bd. III, T. 2, 192.  
Litten, Bericht. d. Heid. Oph. Gesl. 1877, 140.  
Roth, A. f. Oph. LV, 1872. Deutsch. z. f. Chir. I, 1873, 471.  
Treacher Collins.  
Wilbrand, u. Saenger, vol. IV, 1909, 354.

## CHAPTER XI.

### OPTIC NEURITIS AND OPTIC ATROPHY.

**OPTIC NEURITIS.** The study of optic neuritis will be facilitated by recognizing two pathologic conditions which may or may not merge into each other. These are, (1) papilledema or passive, noninflammatory swelling of the nerve head, and (2) true neuritis or active inflammation of the nerve, situated either behind the globe (retrobulbar neuritis) or at the ocular end (intraocular neuritis: papillitis). The term "choked disc" has been arbitrarily applied to any swelling of the nerve head which reaches an elevation of 1 mm. (3.D.) although strictly speaking it should be reserved for cases of noninflammatory edema or papilledema. Nevertheless the term as now employed is clinically convenient.

Two theories are prominent as to the pathogenesis of choked disc, both of which recognize the important anatomic fact that cerebrospinal fluid passes directly from the cranial cavity into the sheaths of the optic nerves: (1) The Manz, Schmidt-Rimpler theory, according to which a high intracranial pressure retards the return lymph current from the nerve sheaths to the brain. This causes distension of the sheaths and forces fluid into the ocular end of the nerve and into the rigid lamina cribrosa, thus strangulating the retinal vessels and the intraocular portion of the nerve (Fig. 139). Parinaud agrees with this theory as regards lymph obstruction, but believes that it acts, not through distension of the sheath but by causing edema of the entire nerve trunk including the intraocular end (Figs. 140-141). (2) The inflammatory theory of Leber assumes that intracranial disease produces toxins which are carried by lymph currents into the optic sheaths where they excite inflammatory reaction in the nerve and disc.

Both the above theories are measurably true, but each is applicable to a different form of neuritis. In choked disc produced experimentally and by brain tumor, the microscope fails to reveal evidence of true inflammation, although, if swelling of the nerve head is excessive, there may be local tissue reaction with round celled infiltration as occurs in the analogous condition of strangulated hernia. On the other hand, in the metastatic and

Fig. 139—Choked Disc. The Sheaths of the Optic Nerve are Distended with Fluid Causing Strangulation of the Retinal Vessels and Intraocular Portion of the Nerve (Manz, Schmidt-Rimpler theory).



Fig. 139.



Fig. 140.

Fig. 140—Choked Disc showing Edema of entire Nerve Trunk, including Intraocular end.

Fig. 141—Choked Disc. Edema of entire Nerve Trunk. Similar to Fig. 140.



Fig. 141.









**Fig. 142.**



**Fig. 143.**

**Fig. 142—Papilledema, or Choked Disc. The Excessive Swelling of the Nerve Head is Non-inflammatory and due to Passive Edema.**

**Fig. 143—Papilledema, or Choked Disc. The same conditions exist as in Fig. 142.**



**Fig. 144.**

**Fig. 144—Choked Disc Following Injury to the Cranium.**



**Fig. 145**

**Fig. 145 — Optic Neuritis in Cerebral Tuberculosis.**

descending neuritis of septic and infectious diseases, in syphilis, tuberculosis, sinus thrombosis, etc., inflammatory infiltration of the nerve is marked although, if only the proximal end of the nerve is involved, the disc may not be swollen. The greatest swelling of the nerve head observable with the ophthalmoscope occurs in papilledema, which is a noninflammatory condition due only to passive congestion and edema (Figs. 142-143).

In the great majority of cases choked disc is obviously due to mechanical compression of the cranial contents by tumors, inflammatory products, depressed bone, excess of cerebrospinal fluid, blood, etc. It appears to be very common after mastoid operations involving the jugular vein or lateral sinus (Stergm. 66). Exceptionally, choked disc is due to meningitis, and to infectious and constitutional disease, especially syphilis; also to injury (Fig. 144), exposure, sunstroke, nasal sinus disease, caisson disease, arteriosclerosis and certain poisons. Other cases appear to be hereditary. Very rarely choked disc accompanies myelitis, disseminated sclerosis and general paresis. In children, optic neuritis usually depends on chronic meningitis or cerebral tuberculosis (Fig. 145). Choked disc is very rare in hydrocephalus externus but is common in acute hydrocephalus internus (serous meningitis) in adults except in those cases attended by nasal discharge of cerebrospinal fluid. Serous meningitis seldom causes choked disc in children in whom excessive brain pressure is relieved by separation of the ununited cranial bones. Uthoff, in 100 cases of syphilis of the nervous system, found choked disc or optic atrophy in 40 per cent. The syphilitic changes consisted of gummatous new formations, 65 per cent; gummatous meningitis (usually basilar), 23 per cent; vascular disease, 8 per cent; syphilitic hydrocephalus internus 2 per cent; double periostitis of orbit 2 per cent. In cerebral hemorrhage 3 per cent of the cases develop choked disc which, according to Uthoff, is always due to percolation of blood from the cranial cavity into the nerve sheath (hematoma of the sheath). The infrequency of choked disc in cerebral hemorrhage may be explained either by rapid recession of high brain pressure or by quick death of the subject.

The full development of a choked disc (papilledema) may occupy weeks or months. Rapid development accompanied by early loss of vision indicates an inflammatory or toxic condition (true neuritis). It is not unusual in papilledema for vision to remain normal for months, showing that the nerve fibers have not been constricted sufficiently to destroy their conductivity. Usually, however, vision is blunted and unless pressure is

relieved it is permanently destroyed. In brain tumor, after full development of papilledema, little ophthalmoscopic change occurs for a long time; months or even a year may pass when, gradually, the swelling subsides, the outlines of the disc reappear and atrophy is established.

**STEREOGRAM 63. PAPILLEDEMA (choked disc) IN BRAIN TUMOR.** Right eyeground of a man aged 37 years. Has double choked discs due to brain tumor. The fellow eye is shown in stereogram 64. The head of the optic nerve forms a mound-like projection rising 1.3 mm. (5.D.) above the level of the fundus. The diameter of the disc apparently is increased owing to extension of edema from the nerve into the neighboring retina. The swollen tissue of the nerve has compressed the retinal vessels and retarded circulation. The arteries are straight and narrowed but, evidently, are conveying more blood to the retina than the obstructed veins can carry away. As a result, the veins are darkened, tortuous and dilated. Tortuosity is extreme in the inferior macular vein which twists like a corkscrew, and also in a vertically ascending vein the convolutions of which rise and fall in a plane perpendicular to the retina. No hemorrhages are seen but serous transudation (retinal edema) has occurred along some of the vessels.

Coloring for a choked disc is supplied by a dark red, intensely hyperemic nerve head overlaid by translucent, edematous nerve fibers. In this nerve the swelling is moderate in amount, the disc has a pale rose color and its surface is delicately veined by the engorged capillaries. In cases attended by excessive swelling, the serous infiltrates may completely obscure the red disc and the nerve head will be very light in color. The light spot observed in the center of this disc is a deep physiologic excavation filled by edematous tissue. Despite the alarming appearance of the fundus, vision is normal.

**DIAGNOSIS.** The broad, light colored, elevated surface exhibited by this disc could be nothing but optic neuritis. In the absence of hemorrhage, venous thrombosis would not be considered in the differential diagnosis. There are no ophthalmoscopic signs which distinguish the choked disc of brain tumor from that caused by other conditions, but in this case the absence of vascular degeneration, hemorrhage and plastic exudation tends to exclude all forms of angiopathic neuritis and indicates a cerebral origin for the swelling.

In all cases of true papilledema it is probable that intracranial pressure is relatively high. The deleterious effect which this exerts upon the optic discs depends upon the unyielding bony walls of the cranial cavity

which vent excessive internal pressure, through the optic foramen and into the intervaginal space around the optic nerve. This is verified in cases of brain tumor where choked discs disappear after intracranial pressure is relieved by trephining the skull.

In the great majority of cases choked disc is caused by tumor of the brain, and of all the symptoms which brain tumor presents, choked disc is most nearly pathognomonic. The ophthalmologist is required not only to discover choked disc but to ascertain its origin; therefore, he should possess a knowledge of its principal cause. In brain tumor papilledema develops in about 80 per cent of all cases. It is said to occur most frequently with cysts and glioma, less often with tuberculous growths. The nature of tubercle is to replace, not displace tissue; therefore, it is possible for a tuberculoma to develop in the brain without increasing the cranial content. The location of a tumor is more influential in causing choked disc than its size; thus, it is almost constant in tumors of the cerebellum and corpora quadragemini, but less frequent in tumors of the corpus callosum and convexity. Choked disc from brain tumor usually is bilateral, but the degree of swelling or stage of progress may vary widely in the two eyes. When unilateral, it may occur in either eye, but is most frequent on the side of the tumor. Tumors situated far forward in the brain are more apt to cause choked disc on the same side than when situated far back, but this is not always the case. In unilateral papilledema the possibility of a growth which involves one nerve must be borne in mind.

The most frequent general symptoms of brain tumor are headache, slow pulse, expulsive vomiting, epileptic seizures, apoplectic attacks and psychic disturbances. The focal symptoms are perversion or loss of function in a spinal, cranial or special nerve. There may be loss of vision without eye lesion, disturbance of speech, inequality of pupils, tremors, localized spasms, choreiform movements, paralysis or contractures. Focal symptoms to be diagnostic of brain tumor must be unilateral and not suddenly developed. Transitory attacks of blindness are a highly significant symptom. Cushing, in 123 cases of brain tumor, found interlacing or inversion of the color fields (dyschromotopsia) in 43 per cent. The cortical centers of vision are in the occipital lobes; consequently, tumors of this region are rich in morbid visual phenomena which strongly suggest, but do not prove, occipital disease. These consist of hemianopsia, hemianopsia for colors with normal fields for white, photopsiæ and visual hallucinations. Mind blindness indicates a lesion of the occipital lobes. Tumors of the hypo-

physis, as in acromegaly, press on the optic chiasm and produce bitemporal hemianopsia. The same phenomenon may be due to hyperdistension of the third ventricle.

Except in syphilitic cases, which may improve under treatment, the course of brain tumor is *steadily progressive*.

STEREOGRAM 64. PAPILLEDEMA (Choked Disc) IN BRAIN TUMOR. Left eyeground of a man 37 years of age. Has brain tumor with bilateral choked discs. The right eye is shown in stereogram 63. In the left eye the papilledema is far more intense than in the right. The nerve head forms an abrupt projection 2.5 mm. (7.5 D.) above the level of the fundus.\* The swelling of the nerve offers great obstruction to the return circulation, as shown by the unusual number of hemorrhages and by the retinal edema, which is more extensive than commonly observed in choked disc. The arteries are small and straight. At the periphery of the fundus the veins form long sinuosities, but toward the disc they become extremely tortuous and dip, with short vertical curves, into the edematous retina until, almost bursting with blood, they force their way into the swollen nerve head. The edematous retina around the nerve presents delicate, radial striations which conform to the course of the nerve fibers. The striate, feathery hemorrhages at the border of the disc and elsewhere are situated in the nerve fiber layer, while the large, dark, rounded extravasations are in the deeper layers of the retina. Vision in this eye is reduced to 10/200. The deterioration is ascribed partly to degenerative changes in the nerve fibers and partly to hemorrhages in the macular region.

DIAGNOSIS. The enormous swelling of this nerve head constitutes an unmistakable choked disc. The question of associated retinal disease always arises in cases of optic neuritis and its presence or absence modifies the diagnosis of the underlying condition. Although, in this case, hemorrhage is abundant, it is far less than occurs in thrombosis of the central vein (Stergm. 46); moreover, neuritis usually is absent in thrombosis and when present is mild in character. In albuminuric retinitis (Stergms. 35-36) the exudates, transudates and hemorrhages occur independently of nerve disease or vascular constriction, while in choked disc they are proportionate to the degree of nerve swelling. The cause of a choked disc is not ascertained from the ophthalmoscopic picture, but from an investigation of the systemic condition.

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\*The height of a choked disc is measured by the difference in refraction between the normal fundus and apex of the nerve.

*Bibliography.*

- Cushing, John Hopkins Hosp. Bul., June 1911. Rep. Section Oph. Am. Med. Assn. 1911, p. 132.  
 Uthoff, Graefe-Saemisch Handb. 2, p. 1043.  
 Manz, Zehender's Monatsbl., Bd. III, 1865. Arch. f. O. XVI, 1, 265.  
 Schmidt, Arch. f. O., XV, 1869, 193.  
 Leber, Graefe Saemisch Handb., Bd. V, 1877. Trans. Int. Cong. 1881.  
 Parinaud, N. & Oliver, vol. III, p. 600.

NEURITIS IN NASAL SINUS DISEASE. Discovery of the relation between diseases of the accessory nasal sinuses and diseases of the eye marked an important advance in medical knowledge. While little is known of the anatomic, histologic or bacterial changes which involve the eye in nasal disease, a mass of clinical data of diagnostic value has accumulated.

The accessory sinuses consist of the maxillary antrum, frontal sinus, ethmoidal sinus and sphenoidal sinus. These cavities frequently are the seat of acute and chronic inflammation, and as they are in contact with the orbit and optic nerve (Fig. 146), the eye usually is more or less disturbed. The inflammatory processes that occur within the cells of the sinuses consist of acute and chronic suppuration, chronic thickening and polypoid degeneration of the mucous membrane, causing distension of the cavities and circulatory disturbance. In neglected cases, caries and necrosis of the bony walls ensue. The serious complications are abscess of the orbit, basilar meningitis, intradural and extradural abscess and thrombosis of the cavernous sinus. The eye is affected chiefly through the optic nerve. The nerve is intimately connected with the cells of the sphenoid and ethmoid, but owing to wide anatomic variations, the relation between nerve and sinuses is inconstant. For example, the nerve may be separated from the sinuses by either thick or thin plates of bone: sometimes it is enclosed within the sinus walls or the optic canal may run entirely within the sphenoidal or ethmoidal cells. The method by which sinus disease affects the optic nerve has not been ascertained, but may be explained either by pressure on the nerve, pressure on the nutrient vessels, local edema or through the action of toxins. In but few cases has infection appeared to reach the nerve by continuity. Necrosis and perforation of bone are not essential to extension of an infectious process from the accessory sinuses to the brain, inasmuch as infection can be conveyed through the venous anastomoses that exist between the mucous membrane and dura mater. A neuritis may develop from secondary disease of the brain, presenting symptoms which vary from those of uncomplicated nasal disease. In



most cases neuritis is unilateral when due to accessory sinus disease, and bilateral when due to brain disease.

**SYMPTOMS.** Owing to the close relationship of the cavities an inflammatory process often is diffuse; special symptoms, however, may indicate localization of disease in certain cavities.

Acute inflammation of the *antrum of Highmore* may be attended by pain about the orbit, lachrymation, unilateral purulent discharge from the nose and, sometimes, disturbed vision. In many cases, however, these symptoms are but slightly developed. If the antrum is occupied by a cyst it may be so distended as to encroach upon the orbit but seldom to the same extent as the other sinuses. When the antrum is occupied by fluid or tumors it does not transilluminate. Opacity can be demonstrated also by means of the X ray. Positive evidence of pus or exudation in the antrum is obtained by puncture.

**FRONTAL SINUS DISEASE** is attended by severe periodic, morning headache and tenderness of the supraorbital and infraorbital nerves. The anterior and nasal walls of the orbit are tender to pressure. Eventually, some part of the sinus wall is apt to bulge into the orbit, usually the upper inner angle. As in antrum disease, both the transilluminator and X ray may be employed to discover opacity in the sinus.

Disease of the *ethmoid* also may produce a cystic tumor which, unless relieved by operation, may open through the skin and form a chronic fistula; or it may perforate into the orbit, the antrum or the nasal cavity. Eye symptoms often appear in disease of the ethmoid.

The *sphenoidal cells* frequently are affected with the ethmoidal.

The close connection of the optic nerve with the ethmoid and sphenoid in more than 40 per cent of posterior sinus inflammations results in visual disturbance. The anatomic variation of these structures, however, is so great that eye phenomena are of little value in localizing the seat of disease, except that in posterior sinusitis eye disturbance is very common and orbital cellulitis is rare, while in frontal sinus disease this order of frequency is reversed. The X ray is employed to photograph the accessory sinuses and when skilfully performed the skiagraphs afford valuable information regarding the presence of exudates in the cells. An occasional early symptom of sinus disease is edema of the lids. Frequently, this is of an ephemeral, recurrent character. It may consist only of slight puffiness most marked on the nasal side of the upper lid, or the entire lid may be swollen. Hard or soft swelling along the orbital ring, due to a low grade periostitis, is common in chronic sinusitis. If the



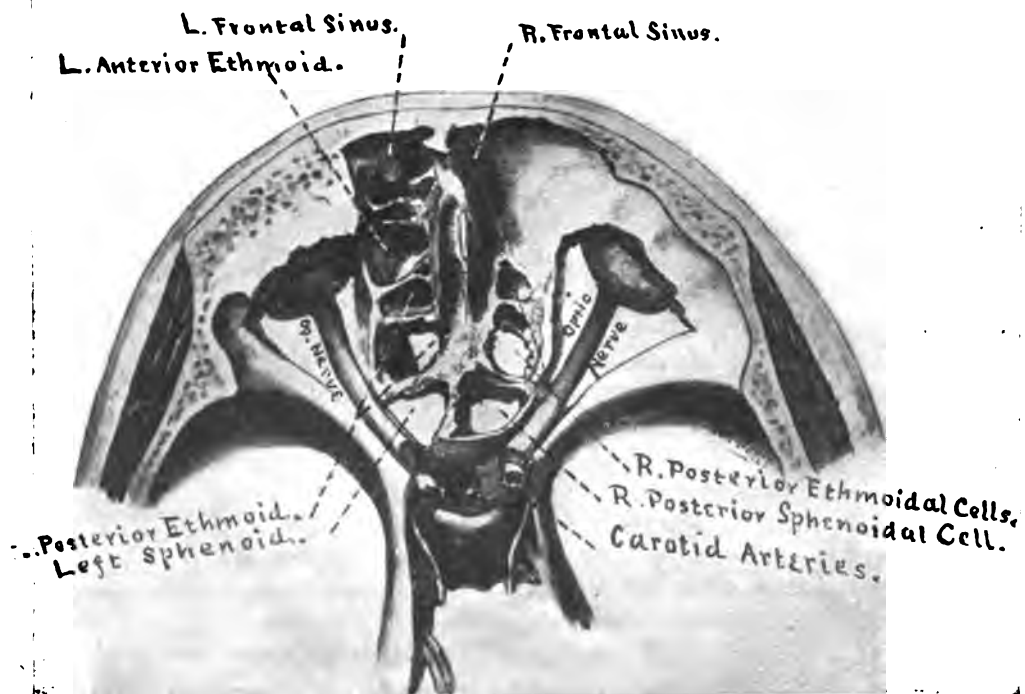


Fig. 146.

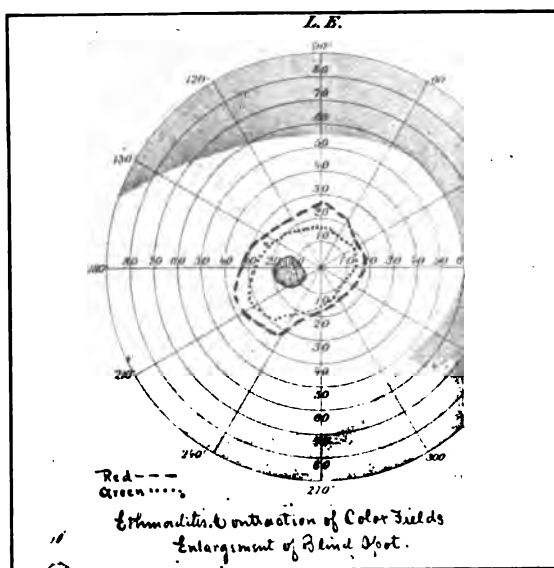


Fig. 147.

distended cells encroach upon the orbit, the eye may be displaced and the patient experience diplopia. Paralysis of the extrinsic ocular muscles may occur in both acute and chronic sinus disease, but is more common in the acute inflammatory type. Pain may be experienced in extreme upward excursion of the eye. The effect of nasal sinus disease upon the optic nerve is of the greatest value in determining the nature of obscure cases. As a rule, only one eye is affected. Bilateral affection of the eyes, especially swelling of the discs, suggests intracranial complications. Changes in the optic disc usually are not marked. They consist of slight marginal obscuration on the nasal side, fullness of the veins and visible perivascular lymph channels on and near the nerve. Even these alterations are present in only a minority of cases. Occasionally, the nerve head is swollen and a mild bulbar neuritis is established. The subjective symptoms indicate a retrobulbar neuritis. Nearly all proved cases show marked contraction of the color fields which may interlace. There is relative central scotoma for colors which in advanced cases may become an absolute scotoma. The normal blind spot is enlarged. The field for form is seldom contracted in the early stages (Fig. 147). While these findings are applicable to the majority of cases, the most varied visual phenomena as hemianopsia, ring scotoma, etc., may develop. Vision may be slightly reduced or it may be completely abolished. Unless the condition is promptly relieved by operation on the sinuses, optic atrophy ensues.

The prognosis as regards vision depends, in a measure, upon the degree of deterioration that exists at the time of operation, but chiefly upon the duration of the process. If optic atrophy is already established the outlook is unfavorable. Harmon Smith got the best visual results from operation in those cases where the sinuses were not markedly diseased.\*

STEREOGRAM 65. Right eyeground of a woman, 50 years of age. Since an attack of influenza six months ago she has suffered from headache. The right eye appears normal. Left eye, vision 20/100. Visual fields show marked contraction for red and green with normal field for form (white). There is mild optic neuritis. The disc is elevated about 0.5 mm. (1.5 D.); on the nasal side the outline is obscure and the adjacent retina is edematous. On and near the disc the vessels are bordered by white lines which are supposed to be dilated perivascular sheaths. All veins are somewhat dilated and one, the superior nasal, is quite tortuous. The arteries are straight.

\*Manhattan Eye, Ear and Throat Hospital Reports, 1912.

The patient was referred to the rhinologic department of the hospital where a diagnosis was made of posterior accessory sinus disease. Operation by Dr. L. Coffin was followed by relief of pain, but vision did not improve and when last seen the optic nerve was atrophic.

**DIAGNOSIS.** Moderate, unilateral swelling of the optic disc should always suggest disease of the accessory nasal sinuses. Neuritis of nasal origin is further distinguished by the subjective symptoms of retrobulbar neuritis, *i. e.*, relative central scotoma for colors and marked contraction of the fields for red and green. The diagnosis should be confirmed by a rhinologic examination.

**THE OPTIC NERVE IN CEREBRAL SINUS THROMBOSIS.** Choked disc and optic neuritis are common events in mastoiditis associated with sinus thrombosis or other intracranial complication. Ruttin, in 30 cases of lateral sinus and jugular operations found 8 in which the optic nerve was affected. This appears to be a high percentage. J. D. Richards examined 100 similar cases and found choked discs in about 10 per cent. The papilledema or neuritis is bilateral in more than half the cases; when unilateral it usually occurs on the same side as the operation. Probably, the proportion of bilateral cases would be increased were the conditions which produce choked disc prolonged. In surgical cases the period at which papilledema develops varies from two days to three months after operation (Ruttin). The majority of cases appear to be simple choked disc due to heightened intracranial pressure or local lymph stasis in the nerve. Others, however, present the symptoms of focal inflammation in the nerve itself, *i. e.*, retrobulbar or intrabulbar neuritis.

The mechanism by which otitic disease produces papilledema has not been determined. Many regard thrombosis of the cavernous sinus as the immediate cause, for the reason that frequently it is associated with choked disc, and also because the return blood from the eye is discharged into this sinus by the ophthalmic vein. As a matter of fact, obstruction in the cavernous sinus does not permanently affect intraocular circulation owing to the free anastomoses which the ophthalmic vein has with both the facial and pterygo-palatine veins. Moreover, complete obstruction of the retinal venous circulation does not cause choked disc but does cause retinal "apoplexy" (Stergm. 46). Cavernous sinus thrombosis can exert no direct influence upon the lymph current in the nerve sheaths, although it may cause obstruction by inducing collateral edema. Uthoff states that one half the cases of cavernous sinus thrombosis show no changes in the nerve. Bartels states that a postoperative choked disc is proof that

the intracranial space has been diminished. Undoubtedly, many cases are due to heightened intracranial pressure produced by effusion of serum. Uthoff considers that choked disc is due in lateral sinus thrombosis to wide ramifications of the clot which extend into the veins from the surface causing meningeal edema, and into the vena Galeni, causing hydrocs of the ventricles. Bartels likewise considers that nearly all cases of postoperative papilledema depend upon serous meningitis and hydrocephalus internus, and when the edema subsides the nerve swelling disappears. He does not regard sepsis alone as causative and cites a case which developed sepsis, nephritis and choked disc. The choked disc soon disappeared, while the other conditions persisted, terminating in endocarditis and death. Septic thrombosis which extends into the orbit does not always produce choked disc, as shown by another case of Bartels in which purulent thrombosis of both cavernous sinuses extended into the orbits causing bilateral phlegmon, but the optic nerves were not affected until late in the disease when pachymeningitis and high brain pressure developed. In this case both cavernous sinuses were absolutely closed, nevertheless the retinal veins were not congested, showing the perfect relief which the normal venous anastomoses afford the ocular circulation. Another similar case showed no choked disc, although a clot extended far into the ophthalmic vein. Bartels discusses another class of cases in which the wound progresses favorably and no intracranial complications are demonstrable, yet the disc becomes choked.

The development of choked disc in postoperative thrombosis does not necessarily mean that the general condition of the patient is worse, in fact, it is of little prognostic import. Thus, it may indicate an increase in intracranial pressure from extension of an abscess or from simple edema. In other cases neuritis may be caused by either local inflammation of the nerve, extension of meningitis to the nerve sheaths or obstruction of the internal optic foramen by edema, pus, exudates, etc.

Intrabulbar and retrobulbar neuritis not attributable to heightened intracranial pressure may occur in postoperative thrombosis. In such cases the neuritis is to be attributed to complications. The following example of this condition occurred in the ear service of Dr. J. F. McKernon.

Woman, 22 years of age. Right postoperative septic thrombosis. The lateral sinus was opened and the jugular was resected. Nineteen days after operation meningitis developed. Three days later she became blind with widely dilated pupils. The right optic nerve gradually became choked, reaching an elevation of nearly 2 mm. Vision in this eye began

to improve before the nerve swelling reached its greatest height. The left eye remained without perception of light for fifty days yet at no time was there any swelling of the disc or congestion of the retinal veins. Both nerves subsequently became white, although vision gradually improved. Figure 148 shows the concentric contraction of the visual fields that existed nine months after operation.

In the above case blindness of both eyes preceded visible nerve changes, differing from papilledema, which does not cause blindness and in which swelling of the nerve antedates whatever visual disturbance occurs. The nature of the pathologic process can only be inferred, although the development of choked disc in the right eye and descending atrophy in the left indicate a retrobulbar focus of inflammation in each nerve, which obstructed the lymph current in one (right) but not in the other. There is little doubt that the conditions were produced by extension of cerebral meningitis to the sheaths of the nerves.

In postoperative neuritis the picture presented by the swollen discs is not always the same. The great majority look like cases of simple obstructive edema. Occasionally, however, the disc presents the appearance of plastic, exudative inflammation (intrabulbar neuritis). The following is a type of such cases.

**STEREOGRAM 66. OPTIC NEURITIS IN POSTOPERATIVE CEREBRAL SINUS THROMBOSIS.\*** Woman 22 years of age. Sinus thrombosis and meningitis followed operation on the right mastoid. Seven days after operation paresis of the right abducens nerve developed and an ophthalmoscopic examination revealed swelling of both optic nerves. The height of the discs at no time exceeded 1 mm. The swollen tissues of the disc had a reddish, congested cast and around the nerve were patches of very dense, white exudate. The veins were moderately congested.

After the neuritis subsided the nerves became very white, presenting a strongly marked picture of postneuritic atrophy.

This case differed from simple edema of the disc in the following respects: the disc was but moderately swollen and the neuritis continued only three weeks, yet postneuritic atrophy resulted with vision of 20/40 and contracted fields (Fig. 149).

In the great majority of postoperative cases swelling of the nerve is due to simple edema and the prognosis as to vision is good. In the

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\*From the ear service of Dr. J. F. McKernon, Manhattan Eye, Ear and Throat Hospital.

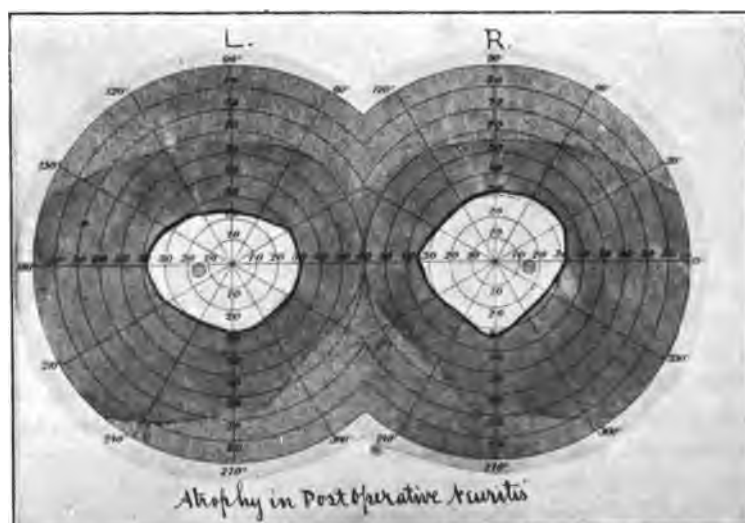


Fig. 148

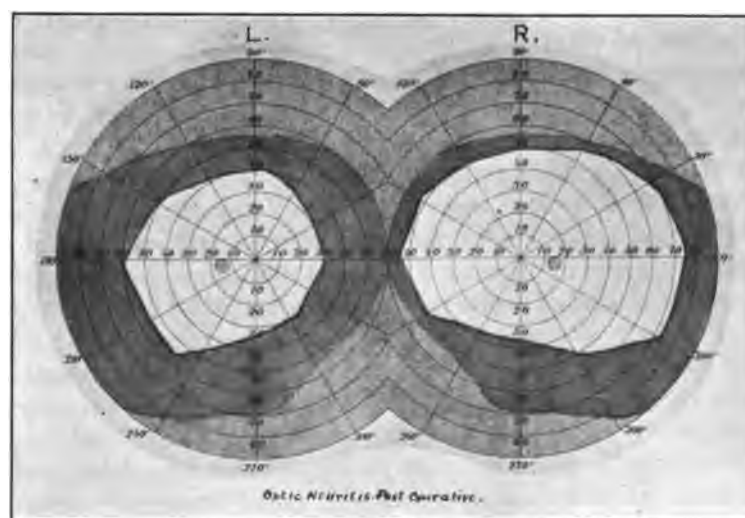


Fig. 149.





rare cases presumably due to inflammation situated in the nerve or nerve sheaths, vision may be impaired but does not appear to be permanently destroyed.

### *Bibliography.*

- Bartels, Neurol. Centralb. 1908, 649; Zeitschr. f. Aug. XXI, Heft. 1. A. f. Aug. LVI, 267.  
 Richards, Personal communication.  
 Ruttin, The Laryngoscope, XXI, Nov. 1911, p. 1051.  
 Uthoff, Graefe-Saemisch Handb. II, Aufl., XI, 703.

**OPTIC ATROPHY.** Ophthalmoscopically, atrophy of the optic nerve is separated into three principal types, *i. e.*, *simple atrophy* in which the nerve fibers disappear by a purely degenerative process; *neuritic* or *post inflammatory atrophy*, in which the nerve fibers are destroyed as the result of inflammation or congestion involving the optic disc, and *retinitic atrophy*, in which destruction of the nerve fibers is secondary to retinitis.

**SIMPLE OPTIC ATROPHY.** In simple atrophy destruction of the nerve fibers may be due to pressure, injury, remote foci of disease or to a general disease of the central nervous system. The best example of simple atrophy is found in those cases where the nerve has been severed or destroyed far behind the eye (Stergm. 25). In certain grave diseases of the nervous system, particularly tabes and multiple sclerosis, optic atrophy may precede by years development of other symptoms of the disease. Inasmuch as these cases are seen early by the ophthalmologist, it behooves him to be familiar with the prominent symptoms of the underlying affection.

**TABES.** Simple optic atrophy (grey atrophy) most frequently is due to tabes and is the first prominent manifestation of that disease in, approximately, 20 per cent of all cases. Both eyes always are affected, but in one atrophy may be delayed. An optic atrophy is recognized as tabetic by two symptoms frequently present in the early stage; these are, (1) the Argyl-Robertson pupillary reaction in which the pupils remain immobile on exposure to light but contract in the acts of convergence and accommodation; (2) loss of patellar tendon reflex. The Argyl-Robertson pupil is diagnostic of cerebral syphilis, usually tabes or paresis.

Other ocular symptoms, such as transitory paralysis of an extra-ocular muscle and, less frequently, contraction or dilation of the pupils, occur in connection with tabes.\* The visual fields of tabetic atrophy are

\*A peculiarity of ocular palsies in tabes is that one muscle of a group may be affected, also that the paralysis may be associated with loss of pupillary light reflex. Permanent paralysis of a single ocular muscle usually is syphilitic.

concentrically contracted (Figs. 150-151). Central scotoma has hitherto been regarded as rare in tabes but Fuchs thinks it is not infrequent. It is yet a question whether such cases are uncomplicated tabes. Contraction of the color fields keeps pace with the field for white and they disappear in physiologic sequence, first green, next red and lastly blue. It is believed that, when tabes commences with optic atrophy, development of incoordination and other symptoms of the disease is delayed. Certainly, in our clinics it is common for patients with tabetic atrophy to remain under observation for years without developing further symptoms of tabes. It appears, however, that the natural course of tabes is extremely slow and ten or even twenty years may intervene between optic atrophy and full development of the disease. Prognosis as to the ultimate result in tabes is extremely unfavorable, but great variations exist in the rate of progress. Deyl gives the following prognostics: The progress will be slow when the nerve is very white and vision remarkably good, also when the fields for white and colors contract uniformly. On the other hand, progress will be rapid when the white fields remain nearly stationary while the color fields rapidly contract, and especially if green is lost. Tabes occurs in middle life. It is far more common in men than in women.

Probably, atrophy of the nerve fibers in tabes is secondary to degeneration of the ganglion cells in the retina although this point has not been definitely decided. The pathologic process appears to be purely degenerative. The nerve fibers lose their myelin sheaths, become varicose and break up, differing in the latter respect from multiple sclerosis, in which disease the continuity of nerve fibers is not destroyed. In the late stage of tabetic atrophy the nerve consists principally of neuroglia and fibrous connective tissue (Fig. 152).

It is stated that at least 95 per cent of tabetics have had syphilis, and tabes is generally regarded as a late manifestation of that disease.

In searching for optic atrophy with the ophthalmoscope, the color of the disc should be studied with a dim light inasmuch as a delicate red tint may exist which a strong light would penetrate and cause the nerve to appear white. In simple optic atrophy the disc at first appears pale, especially on the temporal side; the pallor slowly extends to the nasal side and in course of time the entire disc becomes greyish white. The disc outline becomes sharply defined and the adjacent fundus shows no disturbance. The disc may be depressed but the edges are not undermined; the physiologic markings of the lamina cribrosa become very distinct; the color of the disc changes to a grey or bluish white and the small vessels on the disc disappear but the retinal vessels are not narrowed, at least not until atrophy

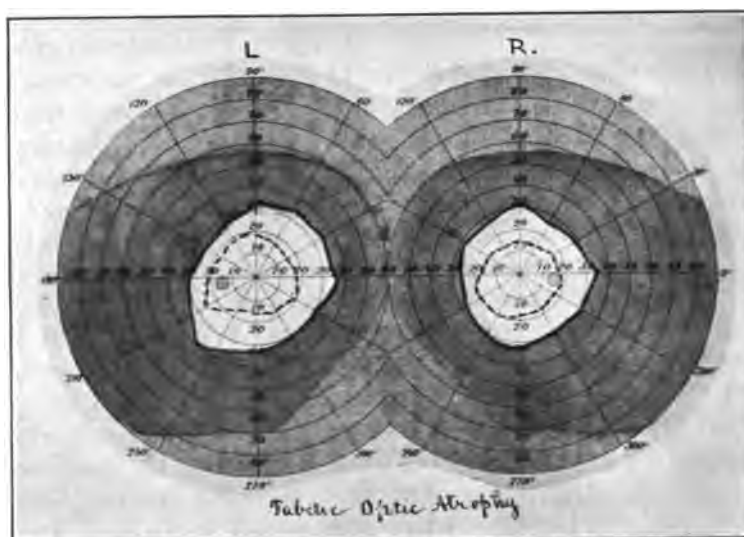


Fig. 150.

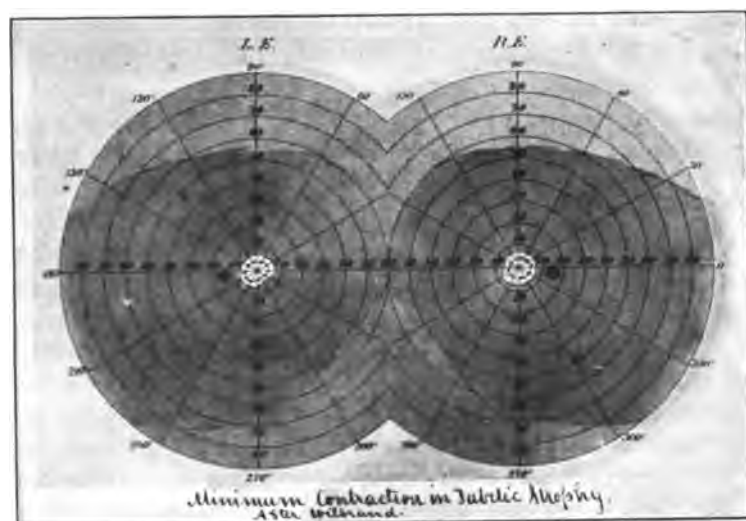


Fig. 151.



is far advanced. If the retinal arteries are diminished in size, atrophy of the nerve is progressing rapidly.

**STEREOGRAM 67. SIMPLE OPTIC ATROPHY IN TABES (Grey Atrophy).** Left fundus oculi of a man aged 51 years. Both eyes affected. He contracted syphilis twenty years ago, for which he underwent prolonged treatment with mercury. Vision has been noticeably failing for the past five years and now is 20/100; there is concentric contraction of the visual fields (Fig. 150); the pupils do not react to light but contract when the patient attempts to read (Argyl-Robertson reaction); patellar tendon reflex abolished; no ataxia. *Ophthalmoscopic Picture:* The optic discs are bluish white in color, regular in outline, sharply defined and slightly depressed. The markings of the lamina cribrosa are very distinct. There is no disturbance of the circumpapillary pigment. The retinal vessels are normal in size and appearance.

**DIAGNOSIS.** This disc presents a typical picture of simple atrophy. Postneuritic atrophy (Stergm. 69) differs, ophthalmoscopically, in that the disc is a dead white, the markings of the lamina cribrosa obscured, the outline of the disc irregular and the retinal vessels narrowed. In retinitic optic atrophy (Stergms. 22-44-53) as distinguished from simple atrophy, the disc has a dirty yellowish color, the cribriform markings are obscured, the retinal vessels are greatly narrowed and the retina is degenerated. The grey atrophic nerve of glaucoma is recognized by the undermined edges of the disc.

While simple optic atrophy is most frequently due to tabes, it occurs also in several other diseases of the nervous system of which the following are the most important.

**MULTIPLE SCLEROSIS.** In multiple sclerosis the brain and cord contain widely distributed plaques of degeneration. The optic nerve, which is a prolongation of the brain, is affected in about one half of all cases. Morphologically, the nerve fibers lose their myelin sheaths, but the axis cylinders, though naked, do not break up as in tabes, neither is their conductivity irrevocably destroyed. The lesions are irregularly distributed and any of the optic nerve fibers may be involved; sometimes only the axial bundle is affected. In optic cases of multiple sclerosis careful examination usually demonstrates disease of both nerves. The degeneration, however, affects the two nerves so unequally that the atrophy is clinically regarded as unilateral in about 75 per cent of the cases. This is in marked contrast to tabetic atrophy which always is bilateral, the two nerves being affected in about equal degrees. Exceptionally, neuritis precedes atrophy, but the majority of cases are atrophic from the first. The ophthalmoscopic pic-

ture in multiple sclerosis accords with the anatomic distribution of the plaques. Thus, the nerve may show atrophy only on the temporal side or a diffuse, incomplete blanching may exist, but the blue, sunken, dead disc of tabes is seldom seen. The visual fields are inconstant. Minute, multiple central scotomata for red and white is a frequent early symptom (Fig. 153). Later, extremely irregular peripheral narrowing occurs. Annular scotoma may develop. In some cases scotomata may develop without visible changes in the nerve. Frequently, the extrinsic eye muscles are paralyzed, the paralysis being incomplete and subject to wide fluctuations. The abducens frequently is affected, also the third but not in all its branches.\* Paralysis of convergence and of lateral conjugate movements is common. A symptom of importance is nystagmus which appears when the eyes are fixed on an object, especially at one side. The pupils may be sluggish but never are affected with myosis or with the Argyll-Robertson reaction as in tabes. The optic nerve may be attacked years before other symptoms of multiple sclerosis are sufficiently pronounced to attract attention.

The disease usually appears between the fifteenth and thirtieth years. It is very rare after thirty-five. It is common in women in whom the fugitive, atypical early symptoms frequently are ascribed to hysteria. In both multiple sclerosis and hysteria uncontrollable, causeless laughter is common. Tremor accompanies all voluntary movements (intention tremor). Other general symptoms are weakness and stiffness in the legs, vertigo, apoplectic-form attacks, headache and disturbance of speech. Sometimes the intelligence suffers. Multiple sclerosis does not lead to complete blindness, and it is characteristic in this disease for the ocular symptoms to subside and temporary recovery ensue. Temporal bleaching of the nerve should always suggest multiple sclerosis, although it occurs also in other conditions such as toxic amblyopia and cerebral syphilis.

STEREOGRAM 68. SIMPLE OPTIC ATROPHY IN MULTIPLE SCLEROSIS.† Right eyeground of a woman 30 years of age with *unilateral* optic atrophy. For the past four years she has been affected with multiple sclerosis. Cannot walk well; has difficulty with speech; memory is very poor; voluntary motions excite tremor; has had transient paralysis of the external ocular muscles; nystagmus develops when she attempts to fix a close object. Right eye, vision is 20/70: irregular, concentric contraction of the visual field. Left eye, vision 20/30: Visual field shows small, scattered central

\* Most deplorable results follow muscle cutting in unrecognized multiple sclerosis and tabes.

† Patient was seen by courtesy of Dr. Edwin G. Zabriskie.



Fig. 152.

Fig. 152—Optic Atrophy of Tabes. Late Stage. The Disc is Cupped. The Nerve Tissue has Disappeared, the Nerve consisting principally of Neuroglia and Connective Tissue.

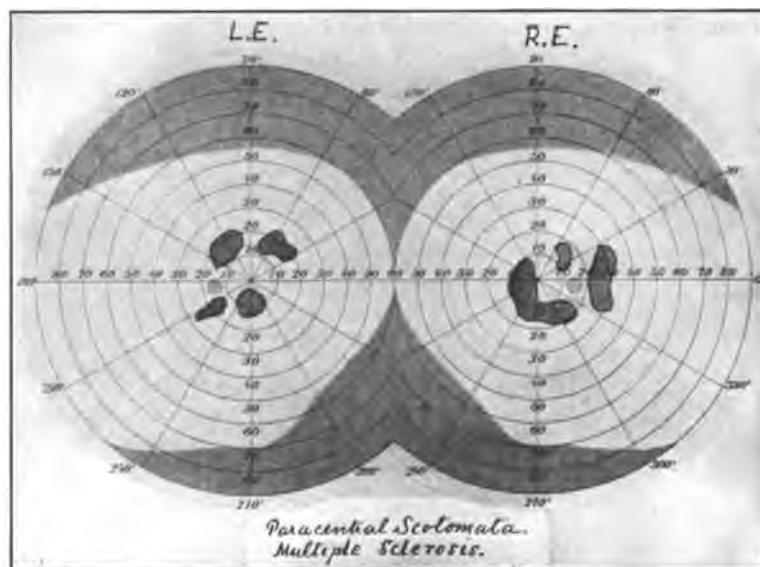


Fig. 153.









Fig 154.  
Oxycephaly, Tower Skull, Turmschädel.

scotomata. This nerve is not ophthalmoscopically atrophic, nevertheless, it is affected. In the right eye the temporal side of the disc is atrophic and decidedly grey in color although the atrophy is not sufficiently advanced to reveal the markings of the lamina cribrosa. On the nasal side the disc presents a normal color. The outline of the nerve is sharply defined and there is no evidence of previous disturbance in the surrounding fundus. The retinal vessels are normal in size and not sclerosed.

**DIAGNOSIS.** In this eye the ophthalmoscopic diagnosis of simple atrophy depends upon the light grey color and sharp outline of the disc, the healthy surrounding fundus and the normal retinal vessels. Limitation of the atrophy to one eye, and especially to the temporal side of the disc, strongly suggests multiple sclerosis as the underlying cause.

**PARESIS.** Paresis is a degenerative disease of the nervous system attended by progressive loss of bodily functions, including a blunted sense of the proprieties and mental enfeeblement terminating in profound dementia. In this affection ocular phenomena are common, especially irregularity and inequality of the pupils, and less frequently the Argyl-Robertson pupillary reaction. Optic atrophy is frequent in uncomplicated paresis and in cases complicated with tabes. It may appear early and occasionally antedates other symptoms.

In *syringomyelia* both choked discs and optic atrophy have been reported, presumably caused by pressure of a distended third ventricle.

Adiposity with optic atrophy usually indicates a tumor of the pituitary body which influences body growth.

*Hereditary Optic Atrophy* is preceded by retrobulbar neuritis, but the disc presents the characteristics of simple atrophy. This peculiar disease attacks only the males of an affected family, but is transmitted through females who themselves escape the affection. As a rule the disease develops during puberty or early manhood.

In *lateral sclerosis of the cord* optic atrophy may appear and present the same symptoms as in tabes.

Optic atrophy may be associated with congenital or acquired deformities of the cranium. In most of these cases the optic foramen has been found to be very small, but it is uncertain whether the atrophy is always due to narrowing of the foramen or both conditions proceed from a common cause. Figure 154 shows a girl four years of age with a "tower skull." She is nearly blind from progressive optic atrophy. In this case it may be assumed that the exophthalmus and optic atrophy are caused by pressure of the confined brain upon the bones which form the roof of the orbit and the optic foramen.

Extreme optic atrophy follows penetrating wounds of the orbit which sever the nerve or cause hemorrhage and compression. Frequently a blow on the edge of the orbit causes a fracture which, extending through the optic foramen, produces injury and atrophy of the nerve (Stergm. 25). In these cases blindness may exist for weeks before the nerve becomes white. The cases reported of optic atrophy in young adults associated with persistent watery discharge from the nose, probably were cases of *hydrocephalus internus*. Simple optic atrophy has been attributed to a great variety of diseases, but it is probable that in most of them atrophy was preceded by papillitis.

**NEURITIC OPTIC ATROPHY.** Atrophy of the optic nerve which has been preceded by neuritis or papilledema is designated neuritic—or post-neuritic atrophy, in contradistinction to simple degeneration of the nerve. A knowledge of the inflammatory processes which lead to optic atrophy is essential. The optic nerves, both in structure and function, are a continuation of the brain, and the brain membranes (dura, arachnoid and pia) ensheath them from the cranial cavity to the eye; consequently, diseases of the brain substance may invade the nerves by direct extension, or meningeal inflammations may spread to the nerve sheaths. The optic nerves may be the seat also of infectious metastases. Basilar extravasations of blood sometimes flow into the optic canals and form a hematoma of the sheath. Inflammation of the sheath is termed *perineuritis*. In this condition the intervaginal space is filled with exudates and the inflammatory processes usually extend to the optic nerve. Perineuritis may be due to syphilis, tuberculosis, meningitis, injuries, periostitis of the orbit, orbital abscess, etc. Inasmuch as the intervaginal spaces of the optic nerve communicate directly with the subdural and subarachnoid spaces of the brain, it follows that heightened intracranial pressure leads to distension of the nerve sheaths and edema of the nerve head (Fig. 139).

Parenchymatous inflammation of the optic nerve frequently is due to syphilis. Gummatous infiltration of the optic tract is most frequent at the chiasm, but any portion of the nerve may be attacked. The histologic changes consist of cellular infiltration and formation of granulation tissue, which is replaced by a cicatrix. In other cases the tissues undergo necrosis, but in either event the nerve fibers are destroyed and atrophy results.

Severe or prolonged swelling of the intraocular end of the nerve, of either inflammatory or noninflammatory origin, is followed by atrophy, but tissue disturbance is greater and atrophy occurs earlier in the inflammatory type. In postneuritic atrophy the nerve fibers perish while neuroglia and newly formed connective tissue cover the disc and enwrap the vessels,

producing an ophthalmoscopic picture quite different from that of simple atrophy. In neuritis and papilledema, infiltration of the tissues extends for a short distance beyond the nerve into the adjacent retina and choroid. Evidence of this remains after the subsidence of swelling, in the form of circumpapillary atrophy and pigment disturbance. The neuroglia on the disc loses its transparency and this, in connection with newly formed connective tissue, may hide the markings of the lamina and fill all depressions on the disc. Neuritic atrophy follows a state of either passive or active venous engorgement, and the veins remain tortuous long after the swelling has subsided and the disc become white. Gradually, however, contraction of the new tissue in the nerve constricts and narrows the retinal vessels which now may exhibit degenerative changes. In the early stage of neuritic atrophy the nerve may retain a reddish grey color but, when the scar tissue is organized and the capillary circulation on the disc abolished, the surface becomes dead-white and the outlines sharply defined. The disc also may appear smaller than normal and irregular in outline. All the manifestations described are more marked after inflammatory neuritis than after papilledema.

The prognosis depends upon the amount of destruction wrought by preceding inflammation and by contraction of cicatricial tissue and, as a rule, is not favorable. Nevertheless, it is better than in tabetic atrophy, which is progressive, while neuritic atrophy, once established, is stationary unless due to syphilis, which may relapse at any time.

STEREOGRAM 69. NEURITIC OPTIC ATROPHY. Right eyeground of a woman 29 years of age. Nine years ago she acquired chancre on the lip from her husband and was treated at the hospital for over a year. Four years later she was ill for a long time and suffered from violent headaches, vertigo and trouble with vision. The Wassermann reaction for syphilis is positive. Central vision is 20/50 in each eye. There is concentric contraction of the visual fields. The ophthalmoscopic picture is that of postneuritic optic atrophy. The white optic disc is rendered very conspicuous by contrast with the surrounding fundus. Its outlines are sharp but slightly irregular. The circumpapillary pigment is superabundant and in some places appears to have been pushed away from the nerve. The disc presents a smooth, dense surface on which all physiologic depressions and markings are obscured. All the retinal vessels are uniformly contracted and the smaller branches have disappeared. Some of the arteries have sclerosed walls, as shown by white bordered blood columns.

DIAGNOSIS. Neuritic atrophy is ophthalmoscopically distinguished by a very white, smooth surface of the nerve as compared with the grey,

cribriform disc of tabes or the yellowish disc of retinitic atrophy. In tabetic atrophy the retinal vessels are normal. In retinitic atrophy the vascular degeneration is greater than in neuritic atrophy.

The following table applies only to typical cases of optic atrophy:

|                                | SIMPLE<br>ATROPHY.                         | NEURITIC<br>ATROPHY.                                               | RETINITIC<br>ATROPHY.                                         | GLAUCOMA.                                                                                                                                       |
|--------------------------------|--------------------------------------------|--------------------------------------------------------------------|---------------------------------------------------------------|-------------------------------------------------------------------------------------------------------------------------------------------------|
| <i>Color of Disc.</i>          | Grey or bluish-white.                      | Dead white.                                                        | Dirty yellowish white.                                        | Greenish grey. Usually darker than simple atrophy. Deep peripheral shadow.                                                                      |
| <i>Outline.</i>                | Sharp, regular, normal size.               | Early stage blurred; later, sharp but irregular; may appear small. | Indistinct. Normal size.                                      | Sharp. May appear enlarged.                                                                                                                     |
| <i>Lamina Cribrosa.</i>        | Physiologic markings very distinct.        | Usually, markings are obscured.                                    | Markings obscured.                                            | Markings very distinct.                                                                                                                         |
| <i>Surface of Disc.</i>        | Normal or slightly depressed.              | May be covered with opaque tissue.                                 | Opaque tissue usually less abundant than in neuritic atrophy. | Excavated to edge of disc. Excavation commences on temporal side.                                                                               |
| <i>Circumpapillary Region.</i> | Unaltered.                                 | Irregularly atrophic. Pigment line shoved away from margin.        | Extensive chorioretinal changes usually present.              | Light colored, depigmented ring, the glaucomatous halo.                                                                                         |
| <i>Retinal Vessels.</i>        | Normal. May be narrowed in very old cases. | Early stage, veins are tortuous; later, all vessels narrowed.      | Vessels narrowed and degenerated.                             | Veins disproportionately filled; arteries narrowed; arteriosclerosis; vessels pushed to nasal side; arterial pulsation if tension is very high. |
| <i>Visual Fields.</i>          | In tabes concentric contraction.           | Irregular or concentric contraction.                               | Depends on form of fundus lesion.                             | Peripheral contraction; begins in nasal field; also, sector defects connected with blind spot.                                                  |
| <i>Course.</i>                 | Progressive.                               | Nonprogressive.                                                    | Nonprogressive unless fundus disease recurs.                  | Progressive.                                                                                                                                    |

**COMPRESSION OF THE OPTIC NERVE IN ARTERIOSCLEROSIS.** In cerebral arteriosclerosis, the mechanical pressure of diseased arteries upon the optic nerve may produce either optic atrophy or neuritis.

Liebrecht designates three points in the course of the nerve particularly subject to arterial pressure. These are:

(1) At the inner opening of the optic canal. This canal, which transmits the optic nerve, consists of a bony portion, 4 mm. in length, and a fibrous portion which extends into the cranial cavity for a distance of 2 mm. The floor of the fibrous extension is formed in part by the carotid artery, while the roof is covered with a fold of dura, the free edge of which is tense and sharp. Between the carotid and roof lies the optic nerve which here is subjected to the vigorous pulsations of the artery. Under normal conditions this creates no disturbance, but when the carotid is greatly dilated and sclerosed, the nerve will be crushed against the free, sharp edge of the roof.

(2) The ophthalmic artery leaves the cranial cavity through the optic canal, where it lies below and to the outer side of the optic nerve. Marked distension and hardening of the artery within the narrow, unyielding walls of this canal always result in compression and deformity of the nerve. According to Liebrecht it is only in the fibrous portion of the canal that the nerve can be injured by pressure of the artery; because, after the artery enters the bony canal, it becomes invested with a dense dural sheath which completely isolates it from the nerve.

(3) Midway between the inner opening of the optic canal and the optic chiasm the nerve passes between the carotid and anterior cerebral arteries. Should these two vessels become sclerosed they will exert undue compression on the nerve.

In addition, sclerosis and aneurismal dilation of the carotids or the arteries in the circle of Willis, may destroy more or less of the optic nerves or chiasm, thereby producing some of the various forms of hemianopsia.

Of the above sites of arterial pressure upon the optic nerve the most frequent is within the optic canal. Here, the pulsations of a sclerosed ophthalmic artery will produce a deep longitudinal sulcus upon the lower surface of the nerve, creating the condition known as "middle furrow formation." In case the artery is markedly dilated and sclerosed, the nerve may thus be split into two lateral parts connected above only by a thin bridge of tissue. The frequent discovery at autopsies of these furrows in the nerve led to the assumption that the papillo-macular bundle of fibers must often be destroyed in this manner and thus produce absolute central



scotoma. In those cases, however, where deep furrows or even splitting of the nerve have been found post-mortem, a history of scotomata during life was lacking. Nevertheless, in most of the cases examined by Otto and by Liebrecht, in which the nerve had been subjected to extreme pressure, microscopic examination showed actual atrophy of some nerve fiber bundles. These findings, if accurate, lead to the inevitable conclusion that the atrophied nerve fibers must have been represented in the retina by blind areas, even though functional disturbance was not recognized by the patient. On examining the visual fields of 80 senile subjects for symptoms of pressure atrophy, Liebrecht found two cases of absolute central scotoma and two of paracentral scotoma. Others exhibited marked contraction of the fields which did not, however, prevent useful vision. These defects were attributed to the mechanical action of sclerotic vessels upon the optic nerve, toxic amblyopia having been excluded. C. S. Bull had under observation for years cases with scotomata and progressive loss of vision which he attributed to nerve atrophy from pressure of sclerosed vessels. Furthermore, he was strongly disposed to regard so-called simple glaucoma as a descending nerve atrophy due to the same cause.

If otherwise unaccountable defects exist in the visual fields of a patient with vascular degeneration, especially if the nerve head is white, a diagnosis of arteriosclerotic pressure atrophy may be warranted. A white disc, however, is not an essential feature of the symptom complex, inasmuch as functional disturbance long precedes bleaching of the disc. This is due to the fact that the point of pressure on the nerve is situated far behind the place of entrance for the retinal vessels where it does not interfere with the circulation and nutrition of the disc. Consequently, there will be no bleaching of the nerve until the defunct fibers have been replaced by connective tissue. In these cases other symptoms of cerebral arteriosclerosis may be present, namely, vertigo on sudden movement of the head or eyes, diffuse headache, irritable disposition, failure of memory, hallucinations of vision and even dementia.

The most meager evidence exists that complete blindness ever is due to pressure of sclerotic arteries upon the optic nerve. These so-called nerves are prolongations of the brain and, like all brain tissue, will endure without loss of function an enormous degree of pressure if gradually applied. It is very doubtful if "middle furrows" in the optic nerve cause appreciable visual disturbance, unless due to extreme and progressive pressure or pressure suddenly exerted. In the few cases of optic atrophy recognized during life and demonstrably due to pressure

from diseased arteries, the anatomical changes were of the coarsest character. Thus, Byron Bramwell described a case of temporal hemianopsia with pale discs which terminated in blindness and mania. Autopsy discovered an enormous aneurism pressing on the optic nerves, chiasm and tracts.

Rampoldi reported the case of a man affected with headache, poor memory, optic atrophy and very low vision. Autopsy showed diffuse arteriosclerosis and a ruptured aneurism of the left internal carotid.

Pechin-Rollin observed a case with optic atrophy and oculo-motor paralysis of the right eye and, in the left eye, ptosis with paralysis of the superior rectus. Post-mortem examination revealed extreme sclerosis of the carotid and ophthalmic arteries. The paralysis was attributed to pressure of the sclerosed carotid upon the nerves in the cavernous sinus.

The histologic changes produced in the optic nerve by extreme pressure of the ophthalmic artery in the optic canal are regarded by Bernheimer, Otto and Liebrecht as those of non-inflammatory pressure atrophy. Certain nerve fibers lying at the bottom of the pressure furrows lose their medullary sheaths and disappear. From this point an ascending and descending atrophy of the affected fibers rapidly ensues. In course of time new blood vessels may develop and be followed by formation of connective tissue. From the microscopic examination of similar cases, Oppenheim and Siemerling conclude that a retrobulbar neuritis is excited at the point of pressure. They found all the evidence of chronic inflammation, *i. e.*, cellular proliferation, increased vascularity and new-formed connective tissue. All microscopic reports which describe atrophic nerve fibers and interstitial sclerosis situated in the periphery of the optic nerve should be regarded with suspicion, inasmuch as the once accepted "peripheral atrophy" is now known to be the normal formation which exists in all optic nerves, even in the new-born.

Instead of resulting in atrophy, pressure of sclerotic arteries on the nerve may cause choked disc. In 1887 Michel reported a case of double choked discs with retention of normal vision and full visual fields. The cause was shown to be distension, tortuosity and hardening of both internal carotids which obstructed the outflow of lymph from the optic canals, causing edema of the nerves and distension of their sheaths. Two similar cases were reported by Stölting. The first exhibited choked disc in one eye and a dirty red nerve head in the other. There was also paresis of both abducens. The clinical diagnosis was brain tumor. At necropsy no tumor was found, but there was extreme sclerosis of the cerebral

arteries. The second case resembled the first including paralysis of the abducens, but no anatomical examination was secured.

Stölting calls attention to the following features presented by these cases which may prove to be characteristic of cerebral arteriosclerosis:

(1) The long duration of the affection, which was ten years in one case and six years in the other.

(2) The optic neuritis showed no tendency to pass into atrophy, which would not be the course pursued by a neuritis dependent on any other chronic disease.

(3) Paralysis of the abducens, probably due to pressure by a sclerosed carotid on the sixth nerve within the cavernous sinus.

Among the above symptoms, oculo-motor paralysis is the most characteristic of cerebral arteriosclerosis and has been observed in several authentic cases.\* The sixth nerve in passing through the cavernous sinus lies in contact with the internal carotid and is the one most frequently affected, but the other ocular nerves may be involved if the artery is greatly distended. In a case reported by Ritter there was complete ophthalmoplegia of the left eye, due to aneurismal dilation and rupture of the ophthalmic artery. The blood clot lay partly within the cranial cavity, partly in the apex of the orbit.

Optic neuritis due to arteriosclerosis is not necessarily of intracranial origin. Sometimes the only ocular manifestation of arteriosclerosis is neuritis due to disease of the vessels distributed to the head of the nerve which here, as in the retina, results in edema, exudation and hemorrhage.

The ophthalmoscopic picture of albuminuric retinitis has been observed in uncomplicated arteriosclerosis. In such cases it is possible that the white retinal exudates may be lymph which has escaped from the perivascular sheaths, as the result of obstruction within the cranial cavity by a distended internal carotid. Purtscher describes a case as *lymphorrhagia* in the fundus in which, following fracture of the skull, the retina was covered with white exudates.

### *Bibliography.*

- Bernheimer, A. f. O., XXXVII, 2, S. 37; 3, S. 36.  
 Bramwell, Byron, Edinb. Med. Jour., Apl. 1887, p. 918.  
 Bull, C. S., Annals of Oph., XIII, 1, Jan. 1904.  
 Fränkel, Virchow. A. f. Path. Anat. 79, S. 509.  
 Greeff, Lehrs, d. Spec. Path. Anat. Auge, S. 416.  
 Halby, Inaug. Dissert., Kiel, 1902.

---

\*Pechin et Rollin, Halby, Fränkel, Stölting, Rigel, Ritter, Michel.

- Liebrecht, A. f. A., XLIV, 1902, 193.  
Michel, A. f. O., XXIII, 1887, 2.  
Moses, Inaug. Dissert. Würtsberg, 1896.  
Oppenheim-Siemerling, Chatite, Annalen, 1897.  
Otto, Untersuch. u. Schnerven Veränderungen bei Arterisklerosie, Berlin, 1893; A. f. A., XLIII, 2.  
Pechin et Rollin, A. d'O. XXIII, 576.  
Purtscher, Bericht u. d. 36 Versaml. d. Oph. Gesel. Heid. 1910, 294.  
Rampoldi, Jahrs. Bericht d. O., 1892, XIII, 327.  
Rigel, Munich Med. Woch., 1899, 1133.  
Ritter, Ref. by Liebrecht, Gesell. f. Natur u. Heilkunde, Dresden, 1887.  
Scultet, Zietert a. Dem. Traite, d. Med. d. Yeux, Paris, 1818.  
Stölting, K. M. f. A., XLIII, 2, 129.  
Von Graefe, Ref. in Wilbrand u. Saenger (Angiecktasie, 1808, 33).

## CHAPTER XII.

### MYOPIA.

In the myopic eye, parallel rays of light come to a focus before they reach the retina, thus producing a blurred image. In the vast majority of cases this is due to elongation of the posterior half of the eyeball and is termed *axial myopia*. Other exceptional and theoretical causes are: increased curvature of the cornea or lens, known as *curvature myopia*; an abnormally high index in the refracting media, called *index myopia*; and forward displacement of the lens.

**AXIAL MYOPIA.** Despite the enormous amount of research work expended upon myopia, neither its etiology nor pathology has been satisfactorily described.

**ETIOLOGY.** The principal reasons which have been advanced to explain myopic elongation of the globe are:

(1) *Subacute inflammation of the choroid and sclera* (sclerochoroiditis posterior). This theory is no longer tenable, the microscope having failed to show evidence of primary inflammation except in cases of syphilis in adults. The hyperemia, exudation and cicatrization found in ordinary myopia are reparative processes which follow stretching and laceration of the membranes. Von Graefe, who advanced this theory, subsequently changed his views.

(2) *Near Work* which requires efforts of accommodation and convergence, was suggested first by Kepler as the cause of myopia. Probably this is causative only in eyes predisposed to the disease, but it is highly injurious in developed cases. A spurious myopia due to spasmodic contraction of the ciliary muscle may occur in an emmetropic or hyperopic eye. Donders showed that near work could not be the only cause of myopia, inasmuch as the highest grades may be found among the illiterate and those who do no close work.

(3) *Compression of the Eye by the Extrinsic Muscles.* When it was established that myopia was attended by distortion of the globe, it was at once suspected that action of the extrinsic muscles was causative, although opinions differed as to the mechanism of their action. Stilling considered that the two obliques, which enwrap the eye, were responsible.

while Arlt believed that muscular pressure on the vortex veins produced congestion which resulted in distension. Other writers have considered action of the internal and external recti as sufficient to elongate the eyeball. Stilling stated that when the roof of the orbit was low the pulley of the superior oblique was so misplaced that this muscle exerted undue pressure upon the posterior segment of the eye, and his measurements tended to prove that myopes have an abnormally low, broad orbit. This theory has not been supported by the anatomic investigations of others. Muscular compression undoubtedly injures an elongated eye, but it is improbable that it could change the form of a normal globe. It is reasonable to suppose, however, that the oblique muscles influence the form of a distensible sclera and determine the position of a posterior staphyloma.

(4) *Short Optic Nerve.* According to this theory the nerve is too short and pulls upon the back of the eye when the posterior pole is rotated outward as in convergence (Hasner, Weiss). Anatomic investigations have failed to establish this view.

(5) *Congenital Weakness of the Sclera at the Posterior Pole.* Schnabel says that congenital anomalies in the choroid and sclera are constant in the posterior staphyloma of Scarpa.

(6) *Contraction of the Ciliary Muscle.* The view that ciliary spasm is in some way connected with the development of low myopia persists, although it is inconceivable that the form of the globe can be influenced by action of the ciliary muscle. Spurious myopia due to contraction of the ciliary muscle often appears to precede the development of nonprogressive myopia, but it is not clear what relationship exists between the two conditions, although it is conceivable that commencing elongation of the globe may excite ciliary spasm.

Little is known regarding the influence of race upon myopia. Parsons states that myopia is very common among the educated natives of India. Stephenson among 918 boys and 231 girls in a London school, found the percentage of myopia in Jewish boys to be six times greater than among the others, and among the Jewish girls it was three and a half times greater. Myopia is more common among women than among men.

Myopia appears to be hereditary but to what extent has not been determined. Pflüger, among 55 families in which one or both parents were myopic, found that 71 per cent of the children had myopia. Schneller, among 1439 scholars of the higher schools found that only 37 per cent of the myopes had myopic parents. Myopia is very rare in new-born infants.

Myopia is divided into two classes which may prove to be distinct affections, although Schnabel asserts the identity of all forms of myopia. The first type has an average range of from 2 to 8 dioptres. It develops coincidentally with school life and becomes stationary about the 18th or 20th year. This is the myopia of adolescence, known also as nonprogressive—and as school myopia. The second type is known as progressive or pernicious myopia and also, from its serious consequences, as malignant myopia.

**STEREOGRAM 70. NONPROGRESSIVE MYOPIA.** Right eyeground of a man aged 30 years. Bookkeeper. With a concave lens of 6.5 dioptres vision is 20/20. His refraction has not changed since he was 18 years of age. The ophthalmoscopic picture is characteristic of benign myopia. The retinal pigment is sufficiently rarefied to reveal the choroidal vessels. The vertical retinal vessels are slightly inclined toward the temporal side and all vessels pursue a straight course. The optic disc is normal in color. On the temporal side it is bordered by a white, crescentic figure the outer border of which is marked by a line of pigment. This figure is known as the myopic crescent or conus.

**DIAGNOSIS.** Myopia is diagnosticated by testing the refraction rather than by the ophthalmoscopic appearance, although the conus and straight retinal vessels in this fundus are characteristic.

By the direct method of ophthalmoscopic examination the myopic fundus is poorly illuminated and the details magnified. If the myopia is high, a direct examination is difficult. With the indirect method, however, the picture becomes very brilliant and sharply defined.

**ANATOMIC CHANGES.** The myopic crescent or conus shown in the preceding stereogram is the most constant ophthalmoscopic feature in myopia, being present in about 70 per cent of all cases. Conus must not be confused with posterior bulging of the sclera (staphyloma of Scarpa), which is a distinct condition. While conus occurs in all degrees of myopia, posterior staphyloma is not to be expected in a myopia of less than 10 D. (Schnabel). The anatomy and significance of the conus are subjects of discussion. In axial myopia, the posterior segment of the sclera, particularly the temporal side, bulges outward, carrying along the attached choroid and widening the aperture through which the nerve enters the eye. At one time it was accepted that the backward excursion of the sclera created the crescent either by inducing atrophy of the choroid or dragging it away from the edge of the disc. This view has been opposed on the ground that, occasionally, a crescent, or what looks like one, is







Fig. 155—Round, or Normally Shaped Globe.

Fig 155.

Fig. 156—Posterior Bulging of the Globe in Myopia, causing its section to be Oval in Shape.



Fig. 156.

seen in emmetropic or hyperopic eyes. Furthermore, it is said that in myopia a crescent may appear at any point around the disc. Stilling considers the crescent to be the white, inner wall of the scleral canal which is exposed because, in myopia, the nerve is drawn obliquely to the temporal side and the scleral foramen enlarged. Schnabel states that the crescent or conus is an imperfect development of the choroid and tissues around the head of the nerve, in fact, a slight coloboma. Despite all theories as to the origin of the myopic crescent, the fact remains that its area may be enormously increased by atrophy of the choroid along its border.

The principal alteration found in the anterior segment of a myopic eye occurs in the ciliary muscle in which the circular fibers (Müller's muscle) are nearly or altogether absent. Müller's muscle is employed in the act of accommodation, but as accommodation is unnecessary in myopia it is supposed that the fibers atrophy from disuse. A thin poorly developed ciliary muscle permits the iris to recede; consequently, the anterior chamber is deeper in myopia than in emmetropia. Myopes have wide pupils, the reason for which is not altogether clear; presumably, it depends on the relation between accommodation and pupillary contraction. The cornea is supposed to be slightly flattened in axial myopia, although this has not been definitely determined. The fundamental anatomic alteration in axial myopia is thinning and distension of the sclera. In 1816, Scarpa described staphyloma of the posterior segment of the eye, but it was Arlt who first recognized its connection with myopia. Myopic bulging nearly always occurs within a circle the center of which is at the outer edge of the optic disc and the periphery at the posterior border of the oblique muscles. In the normal eye the posterior insertion of the inferior oblique is about 4 mm. from the optic nerve; in myopic eyes with posterior staphyloma this distance may be increased to as much as 17 mm. (Schnabel). The globe is thus changed from a rounded (Fig. 155) to an oval form (Fig. 156). In the normal eye the sclera increases in thickness from the equator to the posterior pole; in myopia the reverse is true, and the sclera is thinnest posteriorly, where it may be reduced to one-fourth its normal thickness (Fig. 160). In extreme cases the sclera may stretch until it is thinner than tissue paper. In such cases local areas may yield to intraocular pressure and form abrupt ectasiæ, constituting the *staphyloma verum* of v. Graefe (Fig. 157). These ectasiæ are more apt to occur on the nasal than temporal side of the nerve and may be recognized with the ophthalmoscope. They seldom occur in eyes with less than 20 D. of myopia. One result of myopic distension is to draw the retina

and choroid toward the temporal side and over the nasal edge of the nerve. In high degrees of myopia the retina may thus be dragged to the center of the disc (Fig. 158). The choroid also is supertracted but never extends so far over the nerve as does the movable retina. At the same time traction is exerted on the scleral canal, enlarging the aperture on the temporal side. In the choroid, the vessels atrophy and openings form in the membrane which are repaired by the processes of granulation and cicatrization, as are the lesions of choroiditis. The pigmented retinal epithelium throughout the fundus is rarefied, thus exposing the choroidal vessels. Where the choriocapillaris has disappeared, the retinal epithelium perishes while the neighboring cells proliferate and heap up pigment (Fig. 159). In myopia the pigment shows no tendency to wander into the inner layers of the retina and form branching figures. In areas where the choroid is atrophied the rods and cones degenerate from malnutrition. The inner retinal layers, which are not nourished by the choroid, are unaffected and the retinal vessels pass uninterruptedly across the atrophic spots. Unfortunately, the macula falls within the area of greatest protrusion; consequently, it is especially apt to suffer and pigment proliferation, spots of degeneration and sometimes holes may occur at the fovea. Small degenerated vessels are seen here with the ophthalmoscope as fine, zigzag or branching lines. Macular hemorrhage is not an infrequent disaster in myopia. The vitreous may become liquified and filled with floating opacities. Although the retina is not attached to the choroid, it follows the latter in its backward excursion. Sometimes, however, a fluid vitreous finds its way behind the retina and produces incurable detachment. See detachment of the retina (page 173). Detachment is particularly to be feared in high myopia, although the lower grades are by no means exempt, particularly if subjected to injury.

Subretinal accumulations of fluid, presumably choroidal effusions, are found in myopic eyes at points where the choroid is destroyed. Such fluid appears to be connected with the processes of repair rather than with detachment of the retina (Fig. 160).

**PROGRESSIVE MYOPIA.** The term progressive or pernicious myopia is applied to those cases which are not arrested on completion of body growth, the eye continuing to elongate until the refraction reaches 20 or 30 dioptries. This type differs from the adolescent or acquired myopia in that it develops at an earlier age and may occur among the illiterate and those who never engage in close work. The process of elongation is more rapid and the pathologic changes more profound than in the non-

Fig. 157—Myopia  
with Local Ectasia of  
Sclera, the *Staphyloma*  
*Verum* of Von Graefe



Fig. 157.



Fig. 158—Case of  
high Myopia. The  
Retina and Choroid  
have been drawn over  
the Nasal Edge of the  
Nerve, toward the  
Temporal Margin.

Fig. 158.



progressive form. In pernicious myopia visual acuity always is lowered and, frequently, useful vision is lost from macular lesions.

STEREOGRAM 71. PROGRESSIVE MYOPIA (Pernicious Myopia; Malignant Myopia). Right eyeground of a woman aged 42 years: Cook. The right eye is myopic 20. D.; the left, 18. D. In the right eye, vision with correction is 20/200. Cannot remember that she ever saw well. The fundus presents the ophthalmoscopic picture of pernicious myopia. The retinal pigment has mostly disappeared, leaving the choroidal vessels exposed throughout the entire fundus. The retinal vessels which normally pass vertically upward and downward are dragged to the temporal side and straightened. The disc is surrounded by a wide white ring of atrophic choroid containing a few specks of pigment and traces of choroidal vessels. Evidently, this circumpapillary ring is too extensive to be regarded as only a congenital crescent. On both the nasal and macular sides of the nerve are several atrophic spots or cicatrices bordered by pigment, identical in appearance with those which follow the eruption of exudative choroiditis. The pigment proliferation around the spots in this fundus is rather exuberant for myopia. The softly outlined spots in the upper field are supposed to be gaps in the choroid in process of repair. Several white walled, sclerosed choroidal vessels are seen in which the blood column is reduced to a mere thread. Sclerosis of the choroidal vessels is most advanced near the disc.

DIAGNOSIS. The differentiation of myopia from other conditions depends, of course, upon the refraction of the eye. It must not be forgotten, however, that choroiditis from constitutional causes may occur in a myopic eye and be mistaken for a manifestation of myopia. The choroidal lesions of myopia are comparatively few in number and are limited to the posterior pole, while those of choroiditis are numerous and widely distributed, but favor the equatorial region.

CURVATURE MYOPIA. Myopia may be due to increased curvature of the cornea or surfaces of the lens. Clinically, most if not all cases of corneal myopia occur in the diseased condition which results in conical cornea. Likewise, *lenticular curvature myopia* usually is due to malformation or disease of the lens, which results in anterior or posterior lenticonus. Relaxation or rupture of the suspensory ligament, which releases the lens and permits it to assume a spherical shape also causes myopia. Under the latter conditions, the younger the subject the greater will be the increase in lenticular curvature. Hess has observed myopia develop in phthisical eyes also in iridocyclitis, probably due to relaxation of the zonula. In

the myopia of early senile and diabetic cataract, it is possible that the curvature of the lens is increased (Parsons).

INDEX MYOPIA. Theoretically, myopia may be due to increase in the refractive index of the aqueous, and myopia developing in iridocyclitis, diabetes and jaundice has been attributed to this cause. Experiments, however, tend to disprove that the index of the aqueous in these diseases is ever sufficiently increased to cause appreciable change in the refraction (Hess). *Lenticular index myopia* is common in commencing senile cataract and may amount to several dioptres. The change occurs in the nucleus. Hirschberg states that the myopia of diabetes is due to index changes in the lens and is independent of cataract.

### *Bibliography.*

- Arlt, Krankheite. d. Aug. III, 1856; U. d. Einstel. d. diop. Apparates, 1861.  
 Donders, Anamn. d. Refr. u. Accom., Wein, 1866.  
 Hasner, Parsons, Path. of the Eye, III., 925.  
 Hess, A. f. O. XLVI, 1898.  
 Hirschberg, Deutsch. m. Wochenft., 1887. 1891: Cent. f. A. XIV, 1890.  
 Kepler, Dioptrice, etc., Ausberg, 1611. Ref. in Parsons, P. of the Eye, p. 930.  
 Mauthner, Op. Fehler d. Aug., Wein, 1876.  
 Parsons, Path. of the Eye, III, 908.  
 Pflüger, A. f. O. XXII, 4, 1876; Parsons, IV, p. 1409.  
 Schnabel u. Herrnheiser, Z. f. Heilk., XVI, 1896. Ueber Staph. post., etc. 1895. Norris & Oliver, III, p. 395.  
 Scarpa, Tratt. d. princip. etc., 1816, II, 146.  
 Schneller, A. f. O. XXXII, 3, 1886. C. f. A. XII, 1888. Ber. d. o. G., 1888.  
 Stephenson, Oph. Rev., XI, 1892.  
 Stilling, K. M. f. A., XIII, 1875. A. f. A., XV, 1885. Beitr. d. o. G., 1886. Unt. u. d. Ents. d. Kurtzg., 1887.  
 Von Graefe, A. f. O., 1, 2, 1854.  
 Weiss.







Fig. 159.

Fig. 159—High Myopia with Disappearance of the Choriocapillaris and with Irregular Massing of Pigment.

Fig. 160—Thinning of the Sclera in high Myopia. An Accumulation of Fluid has taken place between the Choroid and Retina, apparently associated with the Process of Repair and not causing Detachment of the Retina, which is in its Normal Position.

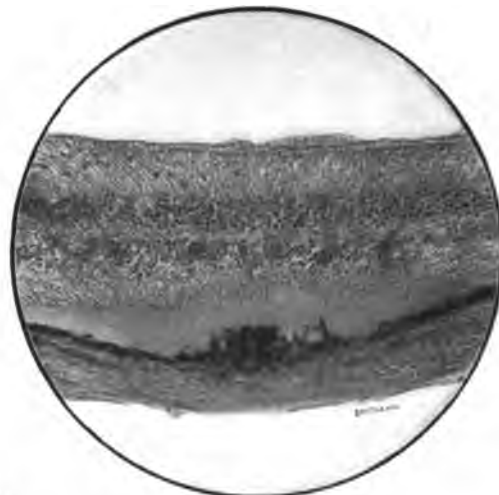


Fig. 160.



Fig. 161

Fig. 161—The Normal Filtration Angle C, cornea; S, canal of Schlemm; P, pectinate ligament; I, iris; C. B, ciliary processes.

## CHAPTER XIII.

### GLAUCOMA.

The essential element of glaucoma is increase in the fluid content of the globe, causing an abnormal rise in intraocular pressure. The normal pressure within the eye which produces *tonus bulbi* or *tension*, is from 18 to 23 mm. Hg. above that of the atmosphere. The maintenance of this condition, which is essential to the integrity of the eye, is largely dependent upon the aqueous humor. The aqueous is produced by the ciliary processes. The fluid enters first the posterior chamber, then passes through the pupil into the anterior chamber. From the anterior chamber the excess of aqueous escapes into the general circulation by filtering through the spaces of Fontana into Schlemm's canal, which is a plexus of veins in the periphery of the cornea (Fig. 161). As these drains are situated in the sinus of the anterior chamber, this region frequently is called the *angle of filtration*.

The tension of the globe being dependent upon the normal production and escape of aqueous, it is evident that any obstacle to its outflow will cause excessive accumulation of fluid in the eye, increase intraocular pressure and establish glaucoma. If high intraocular pressure develops spontaneously without antecedent eye disease or other apparent cause, it is called *primary glaucoma*; on the other hand, if increased tension is due to evident obstruction to the aqueous current by iritic adhesions, displaced lens, etc., it is called *secondary glaucoma*. Primary glaucoma attacks both eyes; secondary glaucoma only the eye which has been previously affected.

**PRIMARY GLAUCOMA.** This form of glaucoma occurs in those who have reached or passed the middle period of life. The course of the disease is fairly uniform and divided into four stages as follows:

(1) *Prodromal Stage.* During this period the patient is subject to transitory attacks of heightened intraocular pressure during which vision is obscured, usually more in one eye than the other; objects seem to be enveloped in a fog and the light of a lamp appears surrounded by a rainbow colored ring. The attack is attended by slight headache or neuralgic pain in the temple or along the side of the nose. On inspection, the

cornea is found to be slightly clouded and there may be some circumcorneal injection. The anterior chamber is shallow. The pupil is moderately dilated and sluggish to the action of light. Where *predisposition* to glaucoma exists, but not otherwise, the attack is determined by anything which causes venous congestion, as mental distress, depression of the heart, dissipation, loss of sleep, constipation, etc. The foggy vision and halos around lights are produced by the "steamy" condition of the cornea. Prodromal attacks usually occur at intervals of weeks. They continue for several hours, but always subside when the patient sleeps, if not before. Exceptionally, the attacks occur with great frequency, sometimes every morning on waking, but pass away in the afternoon. In the intervals the eye is normal except that the power of accommodation is weakened and presbyopia develops in advance of the patient's age. The length of the prodromal period may be difficult to determine, as the initial attacks often are slight and may pass unnoticed. The usual duration is a few weeks or months. It may be prolonged for years, in which case the eye gradually passes into a stage of subacute or chronic glaucoma without having suffered an acute inflammatory attack.

(2) *Glaucoma Evolutum* (Acute Inflammatory Glaucoma; Glaucomatous Crisis). After a longer or shorter prodromal period, the eye suddenly is attacked by an inflammatory glaucoma in which all symptoms of the prodromal period are greatly intensified. These seizures are designated according to their severity as subacute, acute and fulminating glaucoma. Typical inflammatory glaucoma is ushered in by nausea, vomiting, chilly sensations and fever. Excruciating pains develop in the head and eye, often radiating through the ears and teeth. Not infrequently the eye symptoms are so overshadowed by the intense headache as to escape observation and the disease is mistaken for migraine. Eye tension is raised, as may be determined by palpation with the fingers. Vision rapidly fails and may be reduced to perception of shadows. The intense congestive nature of the disease is shown by the external appearance of the eye. The lids are edematous. Intense circumcorneal injection is soon succeeded by bluish, chemotic swelling of the conjunctiva. The cornea is insensitive to the touch and rapidly becomes opaque. The anterior chamber is shallow. The iris is congested, discolored, and the periphery is pushed forward. The pupil is dilated and immobile to light. The pupillary dilation may be unequal, in which case the pupil is oval or eccentrically situated. If seen at the onset of the attack the ophthalmoscope will show pulsation in the retinal arteries, but in a short time opacity of the cornea develops and prevents a view of the fundus.

An acute glaucomatous seizure is precipitated by the same influences as induce prodromal attacks, *i. e.*, anything which produces congestion of the uveal tract. An attack may be due to local causes as contusion of the head or slight injury to the cornea. An operation for glaucoma on one eye may be followed by acute glaucoma in the fellow eye, presumably due to mental depression incident to operation. Dilation of the pupil thickens the root of the iris and crowds it into the angle, thus blocking the outflow of aqueous; therefore, the use of a mydriatic, especially atropine, will induce an attack of glaucoma in an eye *predisposed* to the disease.

An attack of acute glaucoma continues for some days or weeks, when the congestive symptoms gradually subside and the eye again becomes comparatively well; the conjunctival redness and opacity of the cornea disappear and central vision usually improves enough to enable the patient to read. Inflammatory glaucoma, however, always leaves traces of its presence. The vision is never quite so good as before the attack and the nasal field is contracted. The anterior ciliary veins, which emerge through the sclera near the cornea, remain dilated; the anterior chamber is not of normal depth; the iris is thinned and slate colored; the pupil dilated and sluggish; the eye tension is continuously above normal. Such an eye is said to present a glaucomatous aspect (*habitus glaucomatosus*). When the cornea clears sufficiently to permit a view of the fundus, the ophthalmoscope shows the retinal veins somewhat dilated and the arteries of normal width or narrowed. If arterial pulsation does not exist it can be produced by slight pressure on the globe or by use of a mydriatic (which, however, should never be employed in primary glaucoma). The optic disc is cupped and the vessels are transferred to the nasal side. A considerable period of time may elapse before another attack occurs, during which the eye remains quiet. Second and subsequent attacks do not differ from the first, except that they are apt to be less severe. Explosions of the disease recur with increasing frequency, each of which destroys some of the remaining vision until at last the eye becomes blind.

(3) *Glaucoma Absolutum*. In this stage the eye is hard as stone and completely blind. The amount of pain varies; it may be absent or so severe as to require enucleation of the eye. The cornea is transparent but insensitive to the touch. The sclera is thinned and transmits the blue color of the underlying uvea. The anterior ciliary veins are distended and tortuous. The lens and iris are advanced, rendering the anterior chamber very shallow. The pupil is dilated and reacts slowly or not at all to light. The iris becomes greatly atrophied and transillumination shows holes in

its stroma. With the ophthalmoscope the disc is seen to be totally excavated, the retina atrophic and the vessels more or less degenerated. Retinal hemorrhages may occur and are to be regarded as an extremely unfavorable symptom. This stage pursues a very slow course and may continue for years before degeneration is well established.

(4) *Glaucoma Degenerativum*. A glaucomatous eye is classed as degenerative when permanent opacities appear in the cornea and lens. In the cornea parenchymatous opacities develop, while vesicles and hyaloid deposits form on the surface. Opacity of the lens (*cataracta glaucomatosa*) always develops. The glaucomatous cataract is swollen, has a bluish-white color and the surface presents a silk-like luster quite different from the dull, grey, sectoriform opacity of senile cataract. In glaucomatous cataract the eye is blind, hence, operation would be useless. On the other hand, ordinary senile cataract may form in a glaucomatous eye with a fair visual field, in which case extraction may be performed with great benefit to the patient. After a number of years the eye may atrophy or be lost from perforating ulcer of the cornea. Extensive intraocular hemorrhage, spontaneous or following slight injury, may occur and necessitate enucleation. Occasionally, such hemorrhage may cause spontaneous rupture of the eyeball. I have microscopically examined one such case\* (Fig 162).

GLAUCOMA FULMINANS. This term was applied by Von Graefe to extremely violent cases of inflammatory glaucoma in which vision is permanently destroyed in a few hours (Fig. 163).

CHRONIC OR SUBACUTE GLAUCOMA. These terms are employed to distinguish those cases of glaucoma which run their course without at any time manifesting acute congestive or inflammatory symptoms, although the eye becomes injected and painful.

SIMPLE GLAUCOMA. This remarkable type of glaucoma is characterized by almost complete absence of the congestive symptoms that occur in the inflammatory form of the disease. Von Graefe called the condition "amaurosis with glaucomatous excavation of the optic nerve." It usually occurs in middle life but has been seen in the young and also in myopes, who are almost exempt from the inflammatory type. Men and women are affected in about equal numbers. The disease always attacks both eyes, but it often is more advanced in one than in the other. Glaucoma simplex commences insidiously, without pain and exhibits little tendency to remissions or exacerbations. Unless arrested by treatment

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\*From the service of Dr. J. W. Ingalls, Brooklyn Eye and Ear Hospital.

it progresses uninterruptedly to absolute blindness. The eye tension is continuously above normal. Schiötz, using his tonometer, found it higher in simple, than in congestive glaucoma. It is, however, subject to fluctuation; thus, at times increased tension cannot be detected with the fingers but if repeated examinations are made throughout the day hypertension will be discovered. The external appearance of the eye usually is normal, but at certain times, especially if anything has occurred to cause uveal congestion, the cornea will be slightly clouded, central vision a little lowered and a colored halo may appear around lights, as occurs in the prodromal stage of inflammatory glaucoma.

Central vision often remains surprisingly good until late in the disease, but the visual fields undergo great contraction, especially on the nasal side, although sector-like defects and other variations are common. Torpor retinæ often exists, and a field which in a bright light shows little contraction will, when illumination is reduced, present marked defects.

The relationship of simple glaucoma to the inflammatory type has been a subject of controversy and by some it is regarded as a peculiar form of optic atrophy. The identity of the two conditions, however, is indicated by the fact that simple glaucoma may pass into the chronic inflammatory type or an acute inflammatory attack may supervene. Moreover, the essential feature of glaucoma—increased tension—exists in both types. It is probable that, in glaucoma simplex, the very gradual development of high intraocular pressure, enables the ocular circulation to meet the new conditions without inducing congestion.

**HEMORRHAGIC GLAUCOMA.** The term "hemorrhagic glaucoma" is of doubtful utility. It has been applied to a variety of conditions such as secondary glaucoma, which follows intraocular hemorrhage, particularly, hemorrhagic retinitis; also to cases of simple or inflammatory glaucoma occurring in a subject of retinal arteriosclerosis and, hence, particularly liable to intraocular hemorrhage. If employed at all, the name "hemorrhagic glaucoma" should be limited to the latter class which really are cases of primary glaucoma that, owing to the condition of the blood vessels, possess a tendency to hemorrhage.

**OPHTHALMOSCOPIC PICTURE.** In the fundus, the distinguishing feature of glaucoma is the characteristic excavation of the optic nerve. Three types of optic excavation are recognized:

(1) *Physiologic excavation*, which is confined to the center of the disc or, if it touches one side, the margin is not abrupt (Stergms. 5-16-27-40). The lamina cribrosa is in its normal position.

(2) *Atrophic excavation* consisting of a shallow, dish-like excavation of the entire disc without undermined or abrupt edges. There is no recession of the lamina cribrosa (Stergm. 67).

(3) *Glaucomatous excavation* includes the entire disc and the lamina cribrosa is pushed backward (Stergms. 72-73). Simple glaucoma affords the best opportunity to study the changes. In the early stage, the disc is pale and excavation commences on the temporal side but gradually includes the entire disc. The edges of a glaucomatous excavation always are precipitous and in many cases overhanging. The depth of the depression continues to increase throughout the duration of the disease and in advanced cases measures 6 or 7 dioptries with the ophthalmoscope (2. mm.). The nerve undergoes atrophy and acquires a grey or greenish color. The periphery of a glaucomatous disc is shadowed by the walls of the excavation. The apertures in the lamina cribrosa may or may not be visible, depending upon the amount of adventitious tissue in the cup. The retinal vessels are displaced to the nasal side by the early, temporal excavation. Later in the disease they may move temporalward. If tension is high the arteries pulsate spontaneously, but under any conditions pulsation is produced by very slight pressure on the eye. The veins are somewhat dilated and tortuous, while the arteries tend to become narrowed. In advanced cases all the vessels degenerate. Atrophy of the retina and choroid occurs in glaucoma. This appears first around the disc where, in addition to excessive pressure, the nerve fibers are subjected to traction by recession of the nerve. This chorioretinal atrophy, known as the glaucomatous ring, forms a narrow, light colored border around the nerve, gradually increasing in width as the disease advances. The vessels usually follow the contour of the excavation, and when the latter is undermined they dip over the brim and are lost to view until they reappear on the floor of the depression. In some cases the veins cling to the walls of the cup, but the arteries retain their normal positions, while the nerve tissue falls away from them.

STEREOGRAM 72. SIMPLE GLAUCOMA. TOTAL EXCAVATION OF DISC. Left eyeground of a man, 52 years of age. Vision has been gradually deteriorating for the past ten years. Left eye has become totally blind. Right eye, central vision is 20/100. The visual field is contracted, especially on the nasal side (Fig. 164). The eyes have never been painful. Urine normal. Blood pressure 155 mm. Hg. Schiötz tonometer recorded the intraocular pressure as 80 mm. Hg. The anterior chamber is shallow; pupil dilated and very sluggish; cornea, lens and vitreous are transparent. The disc is depressed about 1 mm. (3. D.) and presents



Fig. 162.



Fig. 163.

Fig. 162—Intraocular Hemorrhage in Glaucoma Perforating the Globe through the Cornea and necessitating Enucleation.

Fig. 163—Glaucoma Fulminans. Violent Inflammatory Glaucoma with Rapid Destruction of Vision.

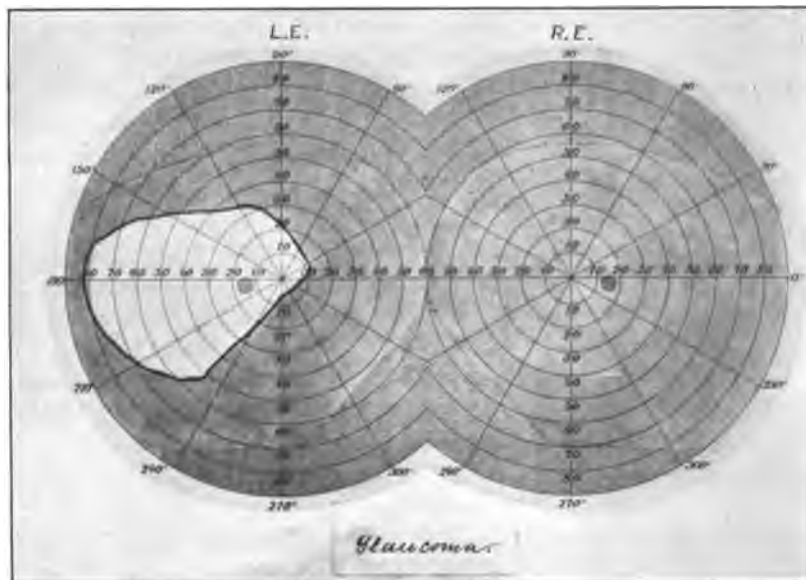


Fig. 164.





the greenish-grey color of optic atrophy. A wide glaucomatous ring surrounds the excavation. The retinal veins are overfull and tortuous. The arteries are narrowed and many of the smaller branches have atrophied. One artery on the nasal side crosses the opening of the pit without sinking until it passes under the excavated edge. Another small artery on the temporal side emerges from the disc, forms a loop and returns into the excavation. This is regarded as an example of the congenital anomaly known as an opticociliary vessel. On the floor of the excavation are two small veins containing short dilations (*Phlebectasiæ*).<sup>\*</sup> Very slight pressure on the globe produces pulsation in both arteries and veins.

Although, in conformity with usage, the term "inflammatory" glaucoma has been retained in this treatise, the name is misleading inasmuch as the attack does not originate in an inflammatory process but is a violent congestion or strangulation of the tissues due to sudden increase in intraocular pressure. The noninflammatory nature of a glaucomatous seizure is indicated by its sudden onset and by its subsidence when pressure is relieved by iridectomy or other measures. It is generally conceded that increased tension is due to some obstacle to the outflow of aqueous, but in primary glaucoma it has not been definitely determined how this originates. If the filtration angle in the anterior chamber is free and unobstructed and the aqueous normal, glaucoma cannot occur. On the other hand, obstruction in this filtration system immediately raises intraocular pressure. Therefore, it is logical to assume that glaucoma is due to blocking these drains, and practical investigators have endeavored to demonstrate how this occurs. Obstruction most frequently is due to approximation of the periphery of the iris to the cornea, and in old cases it becomes permanently fixed in this position (Figs. 165-166). In heightened tension secondary to cyclitis or to retinitis hemorrhagica, the obstacle to filtration occurs at Fontana's spaces and accumulation of fluid in the anterior chamber pushes the iris backward and deepens the chamber (Fig. 102). Primary glaucoma, on the contrary, commences by advancement of the iris. Therefore, the primary cause of obstruction is whatever has advanced the iris, and it is evident that this must be sought for behind the anterior chamber. The following observations of Priestly Smith are of great value and provide a satisfactory explanation for the majority of typical cases of primary glaucoma. Although the eye as a whole ceases

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<sup>\*</sup>For purpose of illustration, details at the bottom of the excavation have been drawn with greater distinctness than they possess when the retinal vessels are ophthalmoscopically in focus.

to enlarge after adult life, the growth of the lens continues, so that at the age of 65 the lens is one-third larger than at 25. Consequently, in course of time the lens becomes too large for the eyeball. The space required to accommodate the enlarged lens is taken from the anterior chamber by advancing the root of the iris and ciliary processes. This explains the way in which the shallow chamber of old age is produced. If, as in hypermetropia, the eye is below the normal size, the continued growth of the lens advances the periphery of the iris beyond the limit of safety. In such eyes swelling of the ciliary processes or dilation of the pupil may push the iris forward in contact with the posterior surface of the cornea and block the angle of filtration. The small eye is especially liable to glaucoma whether or not it is hypermetropic. Clinically, the size of the globe is determined, not by the refraction but by the diameter of the cornea which, as a rule, is less than normal in small eyes.

Fuchs says that the highly vascular ciliary processes may swell and block the narrow circumferential space in an eye predisposed to glaucoma. C. Hess has demonstrated that, in adult life, the ciliary processes become greatly enlarged. The enlargement is progressive, and in the aged bulbus outgrowths form which press forward the iris and tend to induce glaucoma. Not only the processes but the ciliary body itself often becomes cirrhotic and enlarged in old age (Fig. 167). Considerable attention has been directed to the influence of blood pressure (vascular tension) upon the production of glaucoma. In the normal eye tension is kept constant by the aqueous which is regarded as a filtrate from the vessels, hence, intraocular pressure can never exceed the blood pressure in the capillaries, otherwise, aqueous would no longer be produced. Parsons says of the normal eye, "It is most likely that this has the highest capillary pressure of any organ of the human body when at rest." To what extent local capillary pressure varies in glaucoma has not been determined. It has been experimentally demonstrated, however, that intraocular pressure is raised by anything which increases local or general blood pressure in either the arteries or veins. Vascular constriction in distant parts also increases intraocular pressure, thus, stimulation of the abdominal vasoconstrictors drives the blood into the peripheral vessels and a rapid rise of eye tension follows. On the other hand, section of the spinal cord in the cervical regions cuts off the abdominal vasoconstrictors, and the accumulation of blood in the great abdominal vessels which follows causes a decided fall in the tension of the eye. Under normal conditions these variations in eye tension are compensated by changes in the rate of filtration, but in

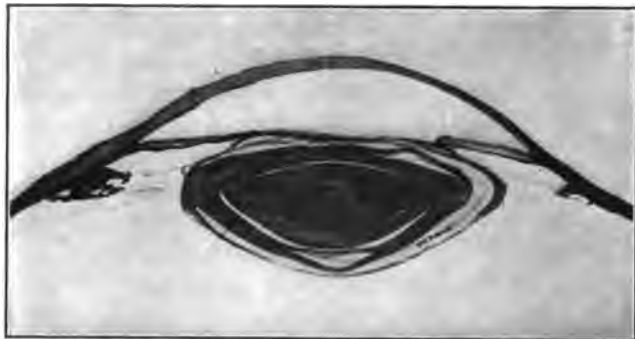


Fig. 165.

Fig. 165—Glaucoma of Long Standing. The Root of the Iris has become Permanently Adherent to the Periphery of the Cornea, Completely Blocking the Filtration Angle.

Fig. 166—The same as 165 but more Highly Magnified. C, cornea; I, root of the Iris; A, filtration angle.



Fig. 166.



Fig. 167.

Fig. 167—Enlargement of Ciliary Body and Ciliary Processes in Advanced Life, Tending to Press the Iris Forward and cause Glaucoma by Narrowing the Filtration Angle.



those suffering from vascular degenerations it is possible that the equilibrium between the arterial and venous systems of the eye is sufficiently disturbed to interfere with circulation in the venous system of Schlemm, which drains the eye. Whatever influence these factors have in creating a predisposition to glaucoma is still undetermined.

Changes in the composition of the aqueous may retard filtration. In the experiments of Priestly Smith, blood serum or ascetic fluid passed very slowly from the anterior chamber. The influence of altered aqueous upon eye tension is clinically observed in subacute cyclitis, in which the aqueous is highly albuminous and loaded with leucocytes. Inasmuch as the spaces of Fontana are filled with leucocytes and albuminous aqueous is filtered with difficulty, intraocular pressure rises. In these cases the anterior chamber is deeper than normal and the angle of filtration is widened. In cyclitis tension does not rise to the same height as in congestive glaucoma, and only in chronic, long protracted cases do the deleterious effects of high pressure ensue.

Pathologists who examine many eyes occasionally encounter cases of glaucoma in which the filtration angle is open and the anterior chamber deepened. Von Hippel published such a case in which the trabeculæ of the pectinate were thickened and the spaces filled with pigment. Baques, in a somewhat similar case following hemorrhagic retinitis, concluded that the hemorrhages in the retina and consequent destruction of retinal tissue, produced irritating chemical substances which acted upon the trabeculæ of the pectinate ligament, causing tissue proliferation which closed the spaces. A case of this kind is described on page 154.

Theoretically, effusions into the vitreous which increase its volume should advance the lens and iris and produce glaucoma. This may occur in intravitreal bleeding if the hemorrhage is sudden and fills the eye. The vitreous, however, appears to accommodate its volume to the space at its disposal and the mere presence of considerable extraneous matter does not raise the tension unless the composition of the aqueous is so changed as to render its filtration difficult. Increase in the volume of the vitreous due to obstructions of fluids or to pathologic changes is regarded by some as a cause of glaucoma, but how this occurs remains to be demonstrated. If pressure in the vitreous exceeds that in the anterior chamber by as much as 1 mm. Hg. the iris is forced against the cornea and the anterior chamber is obliterated.

The etiological importance of choroidal congestion in glaucoma has, undoubtedly, been overrated. It will affect intraocular pressure if ex-

cessive and suddenly developed. Thus, after ligation of *all* the venæ vorticosæ eye tension rises to twice its normal height, but the rise is comparatively slight when but two or three of the veins are tied. After the first abrupt rise tension commences to decline and in a few weeks becomes normal, probably owing to development of collateral anastomoses. The changes which follow ligation of the vorticosæ veins consist of intense congestion, transudation of highly albuminous lymph, opacity of the vitreous and cataract.

A theory has been advanced by Henderson that primary glaucoma is due to fibrosis of the trabeculæ and closure of the spaces in the pectinate. This theory may apply to cases in which the anterior chamber is deepened, as always is the case when the obstruction originates in the filtration spaces.

Other theories held to explain primary glaucoma are choroiditis with excessive serous exudation; anterior iridocyclitis; variations in osmotic pressure in the vitreous; hardening of the sclerotic around the nerve, thereby obstructing the outflow of lymph. It is improbable that obstruction in the lymph channels about the optic nerve and around the venæ vorticosæ can influence eye tension so long as drainage through the anterior chamber is free and unobstructed. Possibly, their closure may exert some influence on the vitreous. Nerve irritation was suggested by Donders as a cause of glaucoma. Eye tension can readily be raised and lowered by irritation or inhibition of nerves, but as this always is accompanied by a corresponding rise and fall in local blood pressure it may be regarded as a compensatory process. High blood pressure is unfavorable in glaucoma, but the influence of arteriosclerosis upon the development of glaucoma has not been determined. It appears reasonable that obstruction might arise from vascular degeneration in the efferent vessels which drain the anterior chamber. Von Graefe first called attention to the influence of heredity in glaucoma and pointed out the curious fact that in *hereditary glaucoma* there is a marked tendency for the disease to anticipate, *i. e.*, to appear in a descendant at an earlier age than in the ancestor. Thus, the prodromata which appeared in a grandparent at 60 appear in a grandchild at 30 or 40. Hereditary glaucoma usually, but not invariably, is exceedingly chronic in its course. Among the constitutional conditions which favor the development of glaucoma *gout* is most prominent.

SECONDARY GLAUCOMA. The course of the aqueous between the ciliary processes and pectinate ligament may be mechanically obstructed by various conditions. The most common are the several forms of iritic adhesions. If the exudate in iritis is markedly fibrinous it adheres to the







Fig. 168.

Fig. 168—Membrane (M) Originating from Inflammation of the Iris (I) and Closing the Pupil (not shown in the figure). *Occlusio Pupillae*.

Fig. 169—Iris Bombé. The Root of the Iris (I) is Adherent to the Cornea (C) and its Pupillary Margin to the Lens (L).



Fig. 169.



Fig. 170.

Fig. 170—Total Posterior Synechia with Advancement of the Lens from Pressure Created in the Posterior Chamber.





Fig. 171.

Fig. 171—Cyst of the Pars Ciliaris Retinae in a Degenerated Eye. The Iris is Adherent to the Cornea, thus Blocking the Filtration Angle and Causing Glaucoma. It is also Adherent by a broad, flat surface, to the Lens, (total posterior synechia).

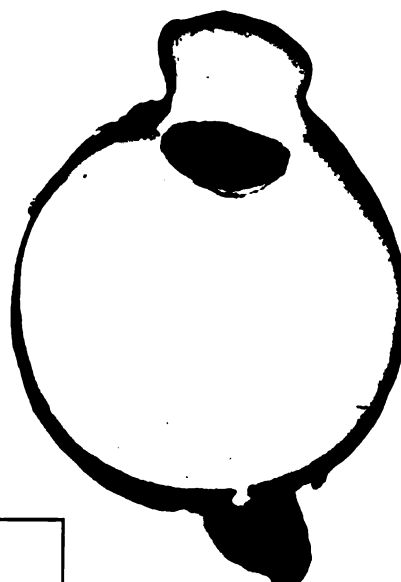


Fig. 172.

Fig. 172—Total Anterior Synechia with Anterior Staphyloma, Causing Secondary Glaucoma. The Optic Disc shows Glaucomatous Cupping.

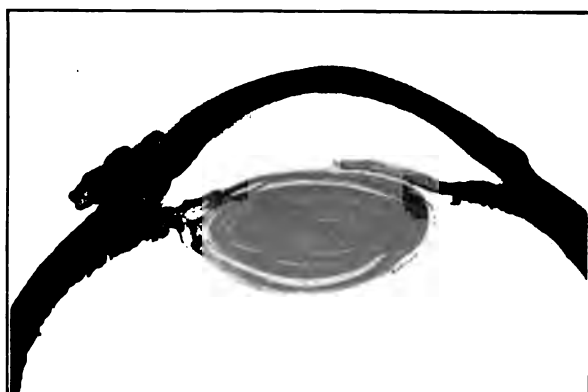


Fig. 173

Fig. 173—Secondary Glaucoma Caused by Lens in Contact with Marginal Corneal Perforation, Blocking the Filtration Angle.



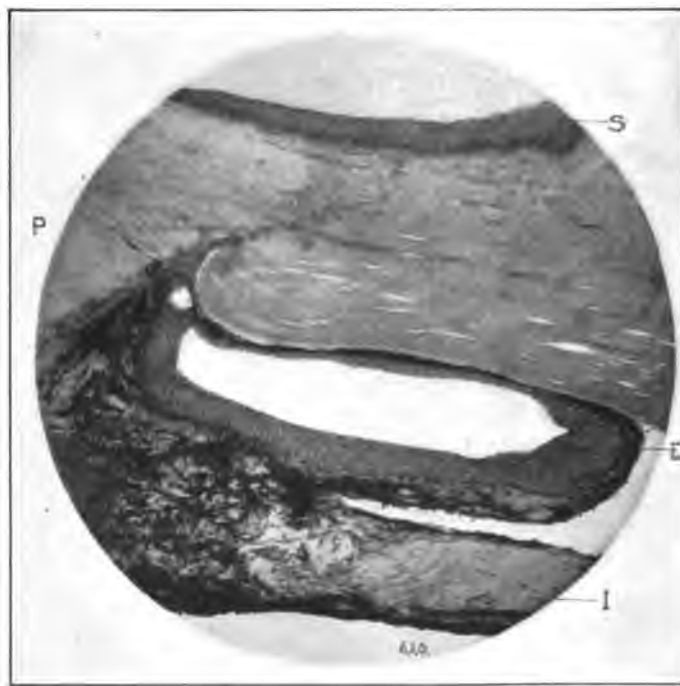


Fig. 174.

Fig. 174—Epithelial Cyst Formation. Continuity of Ingrowing Epithelium has been Severed by Union of Corneal Wound Isolating Cells in Iris. S, surface epithelium; P, corneal cicatrix marking line of perforation; E, epithelial cyst in iris; I, iris.



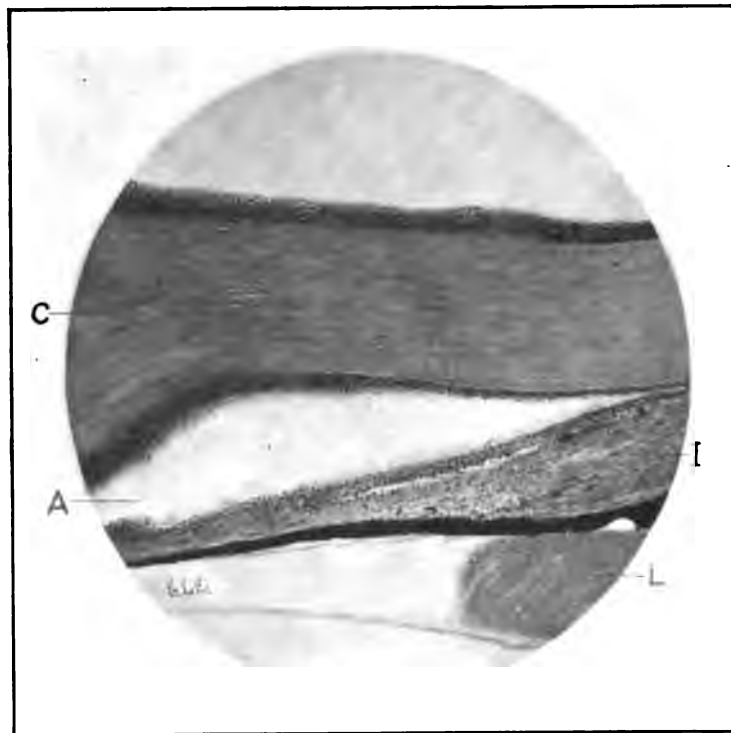


**Fig. 175.**

**Fig.175—Anterior Chamber Obliterated by Inflammatory Adhesions between Iris and Cornea except a Cystic Space (A) lined with Epithelium. C, cicatricial cornea; I, iris; L, lens matter.**







Fig. 176.

Fig. 176—Glaucoma Produced by Epithelium Proliferating into and Blocking the Filtration Angle. S, surface epithelium; C, cornea; B, ciliary body; E, filtration angle.



Fig. 177.

Fig. 177—Spontaneous Dislocation of Lens (L) into the Anterior Chamber, Causing Glaucoma.

anterior surface of the iris and sometimes also to the anterior surface of the lens. In the latter case a membrane which closes the pupil may organize (*occlusio pupillæ*) (Fig. 168). This condition produces glaucoma in which the iris and sometimes also the lens are carried forward against the cornea.

The pupillary margin of the iris may be adherent to the anterior surface of the lens (*posterior annular synechia*). The aqueous being unable to pass through the pupil into the anterior chamber causes the iris to bulge forward producing the condition known as *iris bombé* (Fig. 169).

In *total posterior synechia* broad, flat adhesions exist between the lens and iris. The lens usually is advanced against the cornea by the pressure in the posterior chambers of the eye (Figs. 170-171).

*Total anterior synechia* usually follows a perforating wound or ulcer of the cornea. With occurrence of perforation the aqueous escapes, so that the iris falls in contact with the posterior surface of the cornea and the anterior chamber disappears. Under favorable conditions the perforation closes and the anterior chamber is restored, new aqueous being elaborated. Usually, however, more or less extensive adhesions occur between the iris and cornea. If the pupil is included in the adhesion, secondary glaucoma and bulging of the cornea (*staphyloma*) follow. The protrusion may involve either the whole or only a part of the cornea (Fig. 172). In every instance the protruded portion is composed of opaque cicatricial tissue which develops from the iris. Sometimes the lens is swept into a corneal perforation and remains to block the pupil (Fig. 173).

After perforating wounds of the cornea, the corneal epithelium may proliferate along the wound into the iris and form a cyst (Fig. 174), or it may line the anterior chamber including the pectinate ligament (Figs. 175-176). In either event the angle of filtration becomes blocked and secondary glaucoma develops.

As a result of a blow on the eye, the lens may be driven forward and crush the iris into the angle of filtration where it remains and produces secondary glaucoma (Fig. 132).

Spontaneous dislocation of the lens into the anterior chamber totally obstructs the drainage system of the anterior chamber and results in a pure pressure glaucoma disassociated from inflammation or trauma (Figs. 177-178). Traumatic dislocation of the lens into the anterior chamber also results in glaucoma, but often is associated with other injuries. After perforating wounds of the lens, as after the operation of discission, the lens matter swells and may cause hypertension either by mere increase in size or by loose lens matter blocking the pupil or the angle of filtration.

After cataract extraction about one per cent of the cases develop glaucoma (Knapp). This complication may occur at any time within a year or two after closure of the wound. Risley describes a case of glaucoma which developed in an aphakic eye seventeen years after extraction. After removal of the lens, if a large quantity of cortical matter is left behind it may block the filtration angle or excite sufficient ciliary congestion and swelling to advance the periphery of the iris. Probably, the most frequent cause of hypertension after extraction is incarceration and retention of the lens capsule or iris in the wound. Sometimes vitreous is prolapsed and fills the anterior chamber to the exclusion of the aqueous, in which case it may effectually block drainage. In these cases it is doubtful if the composition of the vitreous is normal. A transparent capsular or inflammatory membrane may completely occlude the pupil and cause glaucoma which abates when the pupillary space is incised with a knife needle. In other cases the pupillary membrane is dense and readily discovered (Fig. 179).

Growths in the ciliary body may advance the iris and mechanically close the angle but in other cases a neoplasm may draw the iris away from the cornea and open the angle, yet glaucoma develops (Fig. 180).

In sarcoma of the choroid, glaucoma appears long before the tumor reaches the ciliary body, but the eye usually does not become glaucomatous until the retina is extensively detached (Fig. 181). Several factors contribute to the development of glaucoma in these cases: (1) Altered vitreous which mechanically pushes forward the lens and periphery of the iris; (2) Irritating effect of tumor juices upon the trabeculæ of the pectinate; (3) Blocking the spaces of Fontana with leucocytes, pigment and debris; (4) Pressure of the tumor on a vortex vein (of doubtful influence).

Evidently, it is impossible to describe all methods by which the filtration angle may be obstructed. The following, however, are not very unusual results of perforating wounds; (1) cystic degeneration of iris and ciliary body (Figs. 171-182); (2) deposit by the endothelial cells of the cornea and iris of a cuticular material resembling Descemet's membrane, in the angle of filtration (Fig. 183).

**MORBID ANATOMY.** The parenchymatous haziness of the cornea in glaucoma is due to disarrangement of its parts. The same effect can be produced in a freshly enucleated eye by compression. In the epithelial layer the cloudiness is caused by edema; the fluid accumulates between the epithelial cells and on the surface of Bowman's membrane. This edema probably is caused by interference with the circumcorneal circulation. In

Fig. 178—Glaucoma Following Spontaneous Dislocation of the Lens into the Anterior Chamber. Although Pressure has caused Ectasia of the Sclera, the Disc does not present a Typical Glaucomatous Cupping. See also Fig. 177.



Fig. 170.

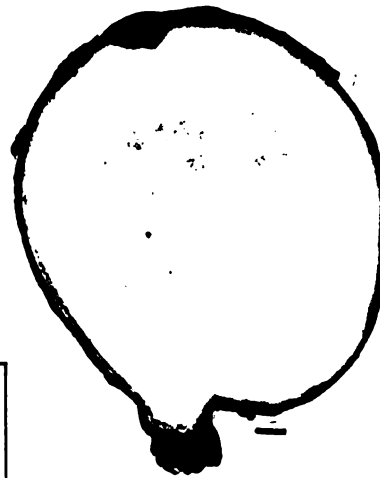


Fig. 179.

Fig. 179—Glaucoma Following Extraction of Cataract and due to a Dense Pupillary Membrane.

Fig. 180—Sarcoma of the Choroid Drawing the Iris away from the Cornea. The Filtration Angle is open. S, sarcoma; F filtration angle.



Fig. 180.



Fig. 181—Sarcoma of the Choroid, with Retinal Detachment, Causing Secondary Glaucoma. The Tumor has not Involved the Ciliary Body.



Fig. 181.

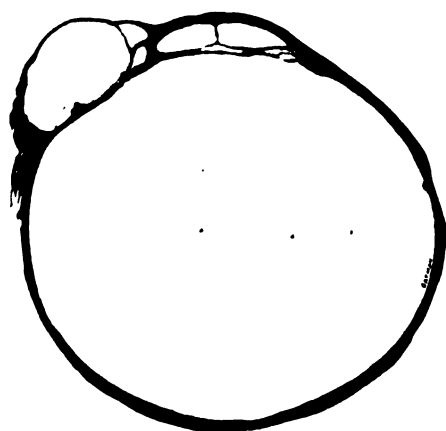


Fig. 182.

Fig. 182—Cystic Degeneration of the Iris and Ciliary Body Blocking the Filtration Angle.

Fig. 183—Filtration Angle Blocked by Membrane Resembling that of Descemet and Derived from the Endothelial Cells of the Cornea and Iris.

- A, filtration angle.
- I, iris.
- D, Descemet's membrane.
- P, newly formed membrane.

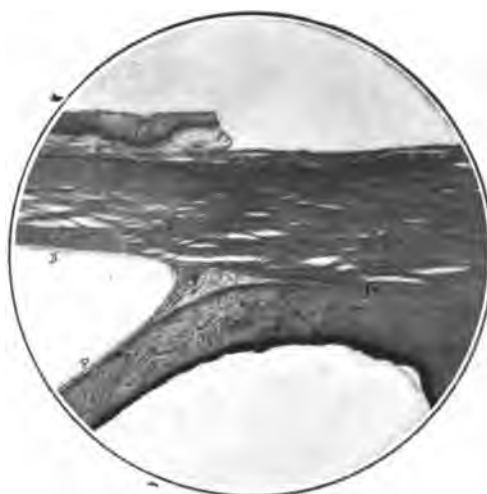


Fig. 183.









Fig. 184.

Fig. 184—Extreme Atrophy of the Iris (I) and Ciliary Body in Old Glaucoma.

Fig. 185 — Ciliary Staphyloma. C, cornea; C. B., ciliary body; L, lens.



Fig. 185.



Fig. 186.

Fig. 186—Intercalary Staphyloma.

nearly all cases submitted to microscopic examination the angle of filtration is obliterated by adhesion of the iris to the cornea. In the early stage the iris is pushed against the cornea by swelling of the ciliary processes but recedes when the attack subsides. In course of time, however, permanent adhesions form between the iris and cornea, thus producing the shallow anterior chamber of glaucoma absolutum (Fig. 165). In the early stage there is inflammatory edema of the iris and ciliary body, but in the late stages the iris becomes too atrophic to manifest inflammatory phenomena. Where the iris is attached to the cornea nothing remains but a line of pigment. In the free portion, the delicate stroma of the iris is replaced by fibrous tissue. The blood vessels are extensively degenerated or entirely obliterated. The muscular fibers undergo hyaloid degeneration. The pigment cells disappear from the stroma, causing the iris to change its color. The posterior pigmented layer is well preserved and as the stroma contracts this layer is dragged forward and over the anterior surface, so that the pupil is surrounded by a black ring constituting the condition known as *ectropion of the uveal pigment*. The ciliary body and iris become flattened from atrophy until in very old cases they are represented by only a line of pigment (Fig. 184). At last the ciliary processes disappear, aqueous is no longer produced and the globe softens. Changes in the choroid correspond to those in other parts of the uvea. At first there is inflammatory edema with exudation of coagulable serum, and in the late stages extreme atrophy. Progressive atrophy of the retina occurs commencing on the temporal side.

The sclera becomes thinner and thinner and at last yields at its weakest part, namely, around the anterior ciliary vessels and at the sclero-corneal junction, where it has been weakened by congestion and inflammation. If the bulging occurs over the ciliary body it is called *ciliary staphyloma* (Fig. 185). If it takes place immediately in front of the ciliary body, it is termed *intercalary staphyloma* (Fig. 186). Equatorial staphyloma occurs around the vena vorticosæ (Fig. 187). Hemorrhage may occur in this region from venous occlusion. Flattening of the cornea results from stretching of the cornea and sclera at their junction.

Excavation of the nerve head in glaucoma is generally attributed solely to increased intraocular pressure, the nerve yielding because the scleral fibers are least numerous at this point. This apparently simple explanation does not altogether conform to the course of the disease or to the pathologic findings, which appear to indicate that the lamina does not yield to excessive pressure until the nerve has been weakened by

atrophy and disappearance of the disc fibers. Although the scleral fibers are least numerous at the lamina, this region is heavily reinforced by the nerve structure and is by no means the least resistant portion of the eye tunics, as is demonstrated by artificially produced pressure from injections of fluid and by cases of pure pressure glaucoma due to dislocation of the lens into the anterior chamber (Figs. 177-178). In estimating the value of observations on this subject, it is important to separate cases in which aqueous obstruction occurred suddenly from those in which it developed slowly or intermittently. In the latter class excavation is extreme. Schnabel regards the excavation as due to neuritic atrophy, which commences by formation of cavities in the nerve tissue which are supposed to fuse and form the glaucomatous excavation. In all the early glaucomas which I have examined microscopically, pit-like depressions were found in which the nerve axons had disappeared, but the neuroglia remained (Fig. 188). Excavation of the disc commences on the temporal side (Figs. 189-190). Continued high pressure soon leads to total excavation of the disc. Optic atrophy precedes or keeps pace with the excavation and in course of time all nerve fibers are destroyed. The glaucomatous cup usually is filled only with vitreous and lined by atrophic nerve fibers (Fig. 191). In some cases, however, it contains a large amount of newly formed connective tissue (Fig. 192). This adventitious tissue may be quite vascular (Fig. 193) and when congested present an ophthalmoscopic picture simulating neuritis. Microscopic sections of glaucomatous eyes frequently show fibrous elevations projecting from the floor of the excavation; they often contain nearly or completely obliterated blood vessels (Fig. 194).

**BUPHTHALMUS** (Infantile Glaucoma; Hydrophthalmus: Ox Eye). Buphthalmus is the glaucoma of infancy. In childhood the sclera is far more distensible than in adult life and when subjected to increased intraocular pressure it yields uniformly. The entire globe is enlarged including the cornea which may form a globular protuberance (keratoglobus) or it may be stretched and flattened. The anterior chamber is very deep. Buphthalmus resembles simple glaucoma in the entire absence of congestive and inflammatory phenomena. In both conditions this may be ascribed to the slow and uniform development of high tension. In the early stages the iris appears normal but gradually it changes color and finally becomes highly atrophic. The lens remains clear until distension and degeneration are well advanced when it becomes cataractous from malnutrition. Subluxation of the lens may occur from stretching and rupture of the zonular fibers. The lens recedes and no longer sup-



Fig. 187.

Fig. 187—Equatorial Staphyloma.



Fig. 188.

Fig. 188—Early Glaucoma with Pit-like Depressions on the Disc, from which the Nerve Axons have Disappeared, the Neuroglia Remaining.

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**Figs. 189 and 190—Early Glaucomatous Cupping of the Disc. The Excavation Commences on the Temporal side.**



**Fig. 189.**

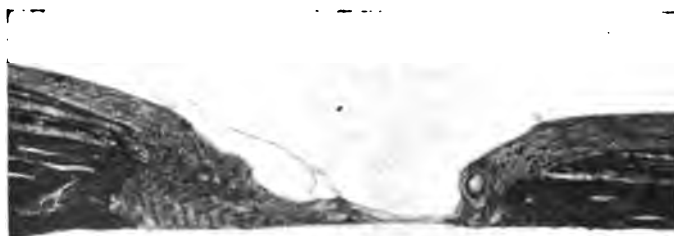


**Fig. 190.**

**Fig. 191 — Glaucomatous Cupping, Later Stage. Complete Excavation. The Optic Nerve is Atrophic.**



**Fig. 191.**



ports the iris so that the latter trembles when the eye is moved (iridodonesis). The retina and choroid undergo atrophy. Although the antero-posterior diameter of the globe is greatly increased, the refraction is not always myopic and the myopia, when present, is not usually of high degree. This is explained by the backward displacement of the lens. Buphthalmus frequently occurs in a family in which a parent and several children are affected by this or some other congenital ocular anomaly.

The obstruction to escape of aqueous is situated in the anterior chamber and appears to be due to arrested development, the iris remaining as in fetal life, in contact with the cornea at the angle of the anterior chamber (Fig. 195). There is also undue persistence of the fetal pectinate ligament. These or other anomalies in the filtration system have been reported in the few cases which have been examined microscopically. It has been experimentally demonstrated that the rate of filtration from buphthalmic eyes is subnormal.

**STEREOGRAM 73. GLAUCOMA WITH ADVANCED ARTERIOSCLEROSIS (So-called Hemorrhagic Glaucoma). EARLY GLAUCOMATOUS EXCAVATION OF NERVE.** Right eyeground of a man 72 years of age. Has general arteriosclerosis. Both eyes are affected with glaucoma simplex. Two weeks ago a "blur" suddenly appeared before the right eye. Vision, 10/200: tension, taken with the finger, is plus. The temporal side of the nerve presents a typical glaucomatous excavation. A light halo in the adjoining fundus marks commencing choroidal atrophy. The excavation extends to the temporal edge of the disc which is slightly undermined. The nasal side of the depression is precipitous, but, has not yet extended to the margin of the nerve. The floor is grey in color and distinctly shows the perforations in the lamina. The main retinal vessels are displaced to the nasal side of the nerve. The ascending and descending retinal arteries are sclerosed as shown where they cross and obscure underlying veins. Numerous small, striate hemorrhages are scattered throughout the macular region and along the supratemporal vessels.

Ten days after the above picture was secured, extensive hemorrhage occurred into the vitreous and blood appeared in the anterior chamber. In two weeks inflammatory glaucoma developed and the eye was enucleated.

**MICROSCOPIC EXAMINATION.** In the nerve, both central vessels were extensively degenerated and the artery was nearly closed by endarteritis proliferans. The retinal vessels presented the usual changes of arterio-



sclerosis. The entire retina was filled with deep and superficial hemorrhages. The optic disc was excavated on the temporal side (Fig. 190). The lamina cribrosa was not displaced.

**DIAGNOSIS.** Acute inflammatory glaucoma is an emergency disease which, if recognized, is readily relieved, while a mistake in diagnosis usually results in destruction of vision. Notwithstanding the vital necessity of recognizing this not uncommon disease, it seldom is diagnosticated by the general practitioner, in fact, a large proportion of glaucoma cases which enter our clinics have had the condition aggravated or even vision destroyed by the use of atropine which has been dropped into the eye for a supposed iritis. During a glaucomatous seizure, the eye condition may be masked by violent pain throughout the distribution of the fifth nerve and the disease is regarded as an attack of megrim. Periodic headache or megrim occurs with considerable regularity from childhood throughout life, although the intervals of attack may grow longer after forty-five. Glaucoma, on the other hand, seldom appears before forty, after which attacks occur with increasing frequency. The aura which precede megrim and enable the patient to foretell a seizure, are absent in glaucoma. The mistake of regarding glaucoma as megrim, however, is less dangerous than that frequently made of diagnosticating and treating the disease as iritis. It is very important to differentiate these two diseases, inasmuch as atropine, which is beneficial in iritis, is destructive in glaucoma. The appearance of the iris is diagnostic; in glaucoma it is pushed forward, especially at the periphery, and the pupil is dilated, oval and immobile to light. In iritis the iris is not advanced, the pupil is contracted, irregular and may be filled with exudates. In glaucoma the iris is free, while in iritis it usually is bound to the lens by synechiæ. The cornea in glaucoma is steamy and insensitive to the touch; in iritis it is clear and sensibility is not affected. In iritis the pain develops slowly and is worse at night; in glaucoma it develops rapidly, is continuous and more severe than in iritis. Vision is lowered in iritis, but may be abolished in acute glaucoma. The deciding symptom in doubtful cases is eye tension, which is normal in uncomplicated iritis,\* but always increased in glaucoma.†

Iridocyclitis, especially of syphilitic origin, may develop violent nocturnal exacerbations attended by vomiting and radiating pains in the supraorbital region. During such an attack eye tension may be increased

\* It is understood that some irritation of the ciliary body always accompanies iritis. The term "uncomplicated iritis" refers to cases presenting no clinical evidence of cyclitis.

† Bear in mind that *secondary* glaucoma may be caused by iritic adhesions.

and the condition so closely simulate inflammatory glaucoma that differentiation is difficult. These cases are recognized as iridocyclitis by—

(1) The history, which in iridocyclitis is that of a slowly developed inflammation.

(2) Contracted pupil and presence of iritic adhesions.

(3) In the secondary glaucoma of iridocyclitis the anterior chamber usually is of normal or increased depth, while in primary glaucoma it always is shallow.

(4) The presence of punctate deposits on the posterior surface of the cornea and on the lens. This is the most important differential symptom. These deposits consist of conglomerations of leucocytes and pigment from the ciliary body which, following the direction of the aqueous current are deposited upon the lens, iris and cornea. As a result of gravitation they tend to settle upon the lower portion of the cornea and should be sought for in this situation by oblique illumination. When the exudate is abundant it fills the sinus of the chamber below, forming a hypopyon. The presence of even a few punctate deposits on the posterior surface of the cornea is proof that the disease is not glaucoma but iridocyclitis.

(5) In cyclitis with increased tension some anesthesia of the cornea may exist, but never is so complete as in inflammatory glaucoma.

The ophthalmoscopic picture of glaucoma is best obtained in the simple and subacute forms of the disease, also in the inflammatory form during the intervals of attack. Early glaucomatous excavation frequently requires differentiation from physiologic excavation of the nerve. In glaucoma the nerve is grey and atrophic and the excavation extends to the very edge of the nerve, which is precipitous or undermined, while in physiologic excavation the depression does not extend to the edge of the nerve. In the oblique, funnel-shaped physiologic excavations (Stergm. 5) the disc gradually slopes from the edge into the cup in a manner quite different from the abrupt edge of a glaucomatous pit.

The ease with which pulsation is induced in a glaucomatous eye by pressure with the finger is diagnostic.

A small coloboma of the nerve may present a superficial resemblance to glaucoma (Stergm. 9), but is readily differentiated by its congenital and non-progressive character and by the otherwise normal condition of the eye.

Intraocular tumors, especially sarcoma of the choroid, induce a secondary inflammatory glaucoma which requires to be diagnosticated from the primary form. Sarcoma is suspected if the eye was blind before the attack of glaucoma and especially if the vision disappeared slowly and

painlessly. Sarcoma occurs in one eye; glaucoma in both eyes, therefore, bilateral glaucoma is evidence against the presence of sarcoma. A history of the characteristic prodromes can usually be obtained in glaucoma. A transilluminator placed over the site of a sarcoma casts a dense shadow on the pupil, but in glaucoma the pupil illuminates perfectly. Hemorrhagic retinitis sometimes is followed by secondary glaucoma in which the history of development closely resembles sarcoma. An important diagnostic point is the appearance of the anterior chamber which may be of normal or increased depth in posthemorrhagic glaucoma.

|                   | ACUTE<br>GLAUCOMA                                                  | ACUTE<br>IRIDOCYCLITIS.                                                                                 | IRITIS.                                                       |
|-------------------|--------------------------------------------------------------------|---------------------------------------------------------------------------------------------------------|---------------------------------------------------------------|
| Development.      | Acute seizure, develops rapidly.                                   | Develops slowly.                                                                                        | Develops slowly.                                              |
| Pain.             | Violent. Radiates through all branches of fifth nerve. Continuous. | Supraorbital and temporal pain. Worse at night.                                                         | Supraorbital and temporal pain. Worse at night.               |
| Eye tension.      | Always increased.                                                  | Frequently increased.                                                                                   | Normal.                                                       |
| Cornea.           | Anesthetic. Parenchymatous opacity.                                | Usually sensitive. Parenchyma clear. Punctate deposits on posterior surface and on lens are diagnostic. | Clear.                                                        |
| Anterior chamber. | Shallow.                                                           | Usually, normal or deepened.                                                                            | Normal.                                                       |
| Iris.             | Congested and discolored. No nodules.                              | Congested and discolored. May contain nodules ( <i>condylomata</i> .)                                   | Congested and discolored. May contain nodules.                |
| Pupil.            | Dilated, oval and immobile.                                        | Contracted. May be bound to lens by synechiæ.                                                           | Contracted. Irregular after dilation with atropine. Synechiæ. |

*Bibliography.*

- Baques, A. f. O. LXVIII, p. 171, 2, Oph. Rev. Aug., 1908, 245.  
 Donders, A. f. O. VIII, 2, 1862; IX, 2, 1863; B. d. o. G., 1864.  
 Fuchs, A. f. O., Bd. XXX, 3. 1884, 125. Text Book, Eleventh Am., 415.  
 Henderson, T. O. S., 1908, 47.  
 Hess, A. of Oph. Jan., 1912, XLI, 29.  
 Parsons, Path. Of The Eye, vol. III, 1047.  
 Risley, Ophthalmology. VI, 4, 1910, 572.  
 Schiötz, Tr. A. Knapp, A. of O. XL, 5, 1911, 518.  
 Schnabel, A. f. A., XXIV, 1892; Wein. m. Woch., 1900; Centb. f. A. 1908, 143.  
 Smith, Priestley, Glaucoma, London, 1879, 1881; Oph. Rev. Apl. 1911.  
 Von Hippel, A. f. O. LII, 3, 1901, 496.  
 Von Graefe, A. f. O. I, 1, 1854; II, 1, 1855; 1857; 1858; 1862; 1869.  
 Goldzieher, C. f. p. A. XXVIII, 1904, 257.  
 Inoyu, O. H. Rep., XVIII, 1910, 24.

## CHAPTER XIV.

### INTRAOCULAR AND ORBITAL TUMORS.

**SARCOMA.** The most frequent intraocular tumor is sarcoma of the choroid. Statistics indicate that it occurs about once in every 1500 cases of eye disease. No age is exempt, but more than half of all cases occur between the 40th and 60th years. In common with other progressive intraocular growths, sarcoma passes through four stages of development:

(1) The preglaucomatous stage, in which the eye is free from pain or irritation. A defect exists in the visual field which, if central, will be recognized by the patient. If the tumor is far forward vision will not be disturbed until collateral detachment of the retina ensues. In small sarcomata the retina usually is in contact with the tumor or separated from it by only a thin layer of fluid in which case the color and form of the neoplasm may be seen. Exceptionally, however, a very small sarcoma produces extensive detachment. In all cases, as the sarcoma increases in size, total detachment results. The usual duration of this stage is from six months to a year; rarely, it continues for years (Fuchs).

(2) The glaucomatous or inflammatory stage, characterized by a progressive, pernicious glaucoma which rapidly becomes absolute. The duration of the second stage is shorter than that of the first.

(3) Stage of extrabulbar extension, which ensues when the sarcoma breaks through the sclera. Perforation usually occurs by extension along blood vessels or along the optic nerve (Fig. 196), very rarely along the ciliary nerves. The opening thus made in the sclera immediately relieves intraocular pressure, and pain disappears. The tumor, released from pressure, grows with increased rapidity. If sarcoma perforates the globe posteriorly, it proliferates luxuriantly in the orbit and causes exophthalmus. If it emerges along an anterior ciliary vessel, sarcoma forms an external granulating mass, usually pigmented.

(4) Usually, the liver is the first organ to receive a metastatic deposit. It is said that, as a rare event, primary sarcoma of the choroid may send out a metastasis before involvement of the sclera. Reports of such cases, however, suggest incomplete microscopic examination.





Fig. 192.



Fig. 193.

Fig. 193—Formation of Vascularized Adventitious Tissue in Glaucomatous Excavation.

Fig. 194—Fibrous Elevation Projecting from the Floor of a Glaucomatous Excavation and Containing a Partially Obliterated Blood-vessel.



Fig. 194.





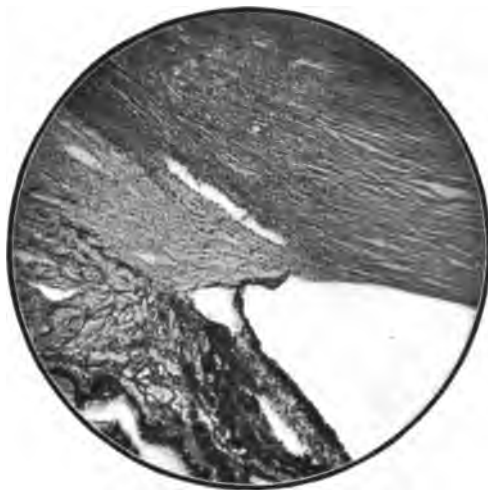


**Fig. 195.**

**Fig. 195—Duphthalmus.** The Iris has Remained in Contact with the Cornea at the Angle of the Anterior Chamber. P, pectinate ligment; S, canal of Schlemm.

**Fig. 196—Sarcoma of the Choroid.** Commencement of Stage of Extra-bulbar Extension.



**Fig. 196.**



**Fig. 197.**

**Fig. 197—Sarcoma of the Choroid, Projecting into the Vitreous.**





Fig. 198.



Fig. 199.

Figs. 198 and 199—Sarcoma of the Choroid Projecting into the Vitreous.



Fig. 200

Fig. 200—Flat Sarcoma of the Choroid.

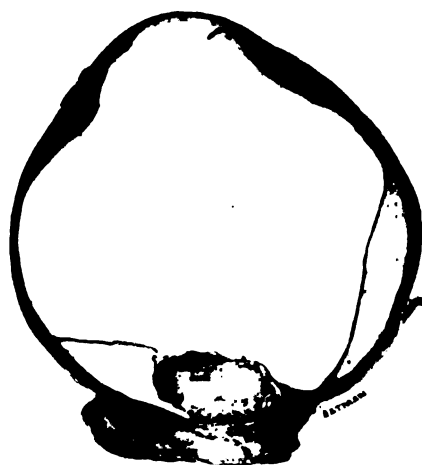


Fig. 201.

Fig. 201—Endothelioma of the Choroid  
(Alveolar Sarcoma).

In exceptional cases the glaucomatous stage of sarcoma is replaced or followed by plastic iridocyclitis, atrophy of the eye and apparent retrocession of the growth. In some of these cases the iridocyclitis may be merely coincidental. In others the sarcoma undergoes necrosis from defective nutrition or hemorrhage and toxins are generated which excite inflammation in the iris and ciliary processes. In all cases, the sarcoma progresses and ultimately becomes actively malignant.

The glaucoma which supervenes upon sarcoma and other intraocular growths appears to be largely mechanical, the thickened vitreous pushing forward the lens and root of the iris, thereby closing the filtration angle in the anterior chamber. Contributing factors are obstruction of the filtration spaces by cellular elements, presence of albumin and fibrin in the aqueous, proliferation of the trabeculæ in the pectinate ligament from the irritating action of toxins in the aqueous and, possibly, congestion due to pressure upon the venæ vorticosæ.\*

In the majority of cases sarcoma of the choroid forms a rounded protuberance situated on a flat, disciform base. This form is determined by environment; thus, in its early stage the neoplasm being compressed between the sclera and lamina vitrea, is low and flat. Soon, the lamina ruptures and the sarcoma grows rapidly into the soft vitreous where it assumes its characteristic mushroom shape, consisting of a globular head in the vitreous, a neck formed by constriction of the lamina vitrea and a flat disc situated beneath the lamina (Figs. 197-198-199). Occasionally, the lamina is not perforated but yields uniformly to a slow-growing sarcoma and a flat discoid tumor results (Fig. 200). A few cases have been reported in which so-called sarcoma appeared as a flat, disseminated infiltration of the choroid; histologically, these growths are endotheliomata (Fig. 201). Sarcoma of the choroid, probably, always is primary. Cases reported as metastatic have not been satisfactorily established. The conditions necessary to establish the diagnosis of metastatic sarcoma of the choroid are the development of sarcoma in an eye *known to be previously healthy* in a person already affected by sarcoma elsewhere. Sarcoma is a solitary growth and it is extremely rare for a secondary deposit to form in the affected eye, differing in this respect from glioma of the retina, me-

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\* See pages 153-4 and 260.

tastatic carcinoma of the choroid and conglomerate tubercle. I have seen one case of local metastasis in flat sarcoma (endothelioma) of the ciliary body.

A woman 40 years of age entered the clinic with no perception of light in the right eye, high intraocular pressure and cataractous lens. The history and examination indicated intraocular tumor and the eye was enucleated. Microscopic examination showed the ciliary body entirely replaced by a typical flat sarcoma (endothelioma, sarcoma carcinomatodes). On the optic nerve was a similar growth which formed a globular protuberance in the vitreous and also extended into the orbit (Fig. 201). These ciliary tumors always are of extremely slow growth, while vitreous and orbital deposits develop very rapidly; therefore, the ciliary tumor is regarded as the primary tumor, particularly, as it corresponded in all respects to other reported cases of flat sarcoma. The globe was sectioned serially and no connection existed between the two deposits.

A few cases have been reported in which with sarcoma in one eye, a plastic iridocyclitis, supposed to be sympathetic, has developed in the other. The assumption that these are cases of sympathetic ophthalmitis does not appear to be justified, inasmuch as iridocyclitis is a common disease and no more frequent among those affected with sarcoma than among others.

Sarcoma of the choroid, like sarcoma in other regions, consists of an embryonic type of connective tissue cells thickly embedded in a scanty, intercellular substance. The predominating type of cell in a given tumor may be spindle-shaped or round. Frequently, the two types are mixed. The occurrence of true giant-celled sarcoma in the choroid has not been established. Sarcoma cells vary greatly in size. Spindle celled tumors are by far the most common in the choroid. The spindle cells have large oval nuclei and frequently their extremities bifurcate. They are arranged in closely packed, interlacing bundles (Fig. 202). In round celled sarcoma the cells often are irregular in shape. Sarcomata usually are very vascular. The newly developed blood vessels consist of thin, endothelial lined tubes and channels, embedded in tumor cells. As a rule round celled sarcomata are more vascular than the spindle-celled type. A sarcoma may consist almost wholly of blood channels or tubes composed of cells. Such growths are termed angiosarcomata and possess a gross microscopic resemblance to glioma of the retina (Fig. 203). Round-celled sarcomata are more rapidly malignant than spindle-celled tumors.

Fig. 202 — Spindle  
celled Sarcoma of  
the Choroid.



Fig. 202.



Fig. 203 — Angio-Sarcoma  
of the Choroid.

Fig. 203.

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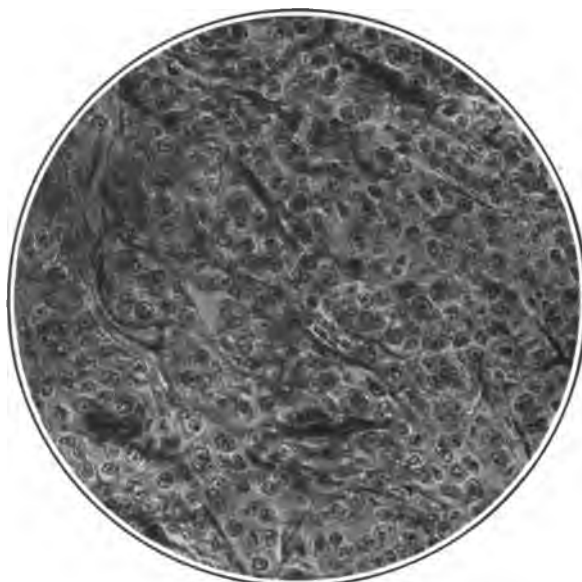


Fig. 204.

Fig. 204—Alveolar Sarcoma of the Choroid (endothelioma).

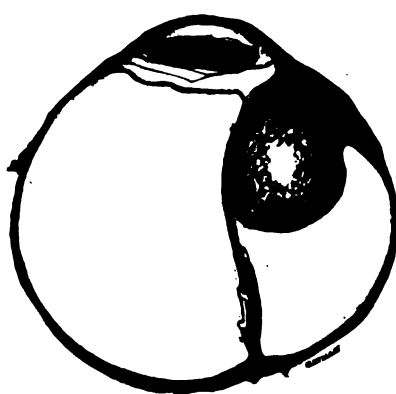


Fig. 205.



Fig. 206.

Fig. 205—Tumor of Choroid and Ciliary Body, which on section (Fig. 206) proved to be a Perithelioma. Note the Rosette-like Arrangement of the Cells, resembling Glioma.

Alveolar sarcoma (endothelioma, sarcoma carcinomatodes) appears in the uvea as the flat, diffuse sarcoma of the choroid and as the ring sarcoma of the ciliary body (Fig. 201). Probably, these growths arise from the endothelium of the lymph clefts and from the perivascular lymph channels. Histologically they are composed of large endothelial cells in a distinct reticulum (Fig. 204). These growths differ from sarcoma in origin, type of cell, structure, tendency to local metastasis and relatively low malignancy. They are classed with sarcoma, however, because both develop from mesoblastic tissue.

A rare type of angiosarcoma or better, endothelioma, is termed perithelioma. This growth is distinguished from other endotheliomata by the type and arrangement of its cells. I have seen but one example of these growths\* (Fig. 205). The cells are of columnar or oval endothelial type and possess abundant protoplasm and rounded or elongated nuclei. They grow in a radiate manner around the blood channels in immediate contact with the endothelial membrane (Fig. 206). Such vessels, when cut transversely, appear as cellular rosettes resembling those found in glioma of the retina. These tumors are supposed to develop from the perithelium of the blood vessels. The perithelium is a single layer of flat endotheloid cells situated between the adventitia and perivascular lymph space, and exists only in certain organs.

A convenient clinical classification of sarcoma of the choroid as melanosarcoma and leucosarcoma is based on the color or degree of pigmentation which the tumor microscopically presents. Those classed as leucosarcoma usually, if not invariably, contain some pigment granules visible under the microscope. Fuchs estimates that 88 per cent should be classed as melanotic. The pigment occurs both in the cellular protoplasm and isolated (Fig. 207). Much controversy exists as to which layer of the choroid is the starting point of sarcoma. Probably, Shieck is correct in his statement that sarcoma of the choroid is no exception to the rule that sarcoma may develop wherever connective tissue normally exists and therefore may arise from the choriocapillaris as well as from the other layers.

The mortality from sarcoma of the choroid could be reduced if instead of preserving enucleated specimens for microscopic demonstration they were always subjected to rigorous microscopic examination for evi-

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\*The eye was microscopically examined for Dr. Jerome B. Thomas.

dence of extraocular extension. The early migration of tumor cells along the optic nerve or the perforating vessels is invisible to the naked eye but may be discovered with the microscope and prompt removal of the orbital contents may be expected to effect a cure (Fig. 208). On the other hand, if left undisturbed, recurrence and death are almost certain.

STEREOGRAM 74. SARCOMA OF THE CHOROID (Leucosarcoma).\* Right fundus oculi of a healthy farmer, 44 years of age. Left eye normal. In the right eye vision has gradually been disappearing for the past six months. Obscuration began on the temporal side and slowly extended like a black curtain being drawn across the visual field. There has been no pain in the eye. *Present condition:* Externally, the eye appears healthy. Tension normal. The ophthalmoscope reveals a circumscribed, globular tumor at the posterior pole, close to the nerve, on the nasal side. This is diagnosticated as sarcoma from its form, immobility on movement of the eye and light flesh color. Several large vessels cross its surface which, from their direction and method of division are supposed to be retinal. No adventitious vessels are seen. A small, simple detachment of the retina adjoins the tumor below, recognized by a slight tremor of the surface on sudden movement of the eye, and also by its color.

The eye was enucleated and microscopic examination showed that the tumor seen with the ophthalmoscope was a small leucosarcoma (Fig. 209).

DIAGNOSIS. The diagnosis of sarcoma of the choroid is comparatively easy when, as in the above case, there is but little collateral detachment of the retina. It becomes difficult, however, when the tumor is small and the detachment total. In every detachment the possible presence of intraocular tumor should be considered. Even traumatic detachments are not beyond suspicion, inasmuch as slight injury suffices to produce detachment when a tumor exists in the eye. The ophthalmoscopic picture of sarcomatous detachment varies. Where the retina lies in contact with the tumor or the layer of fluid between tumor and retina is thin, the surface will be smooth and the vessels will not be crinkled as they are in simple detachment. If, at the same time, the retina retains its transparency the color and form of the underlying tumor will be visible and anomalous vessels belonging to the growth sometimes are seen. Partial sarcomatous detachment soon becomes total; the retina becomes opaque and the pres-

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\* Patient was under the care of Dr. David Webster, Manhattan Eye, Ear and Throat Hospital.

**Fig. 207—Melano-Sarcoma of the Choroid, Showing Pigment in the Cells and Isolated.**



**Fig. 207.**



**Fig. 208.**

**Fig. 208—Sarcoma of the Choroid. The Tumor (S) has Invaded the Optic Nerve and Passed Through the Sclera.**

**Fig. 209—Small Leuco-Sarcoma at the Posterior Pole.**



**Fig. 209.**

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ence of intraocular tumor can no longer be determined with the ophthalmoscope. At this stage the diagnosis is made from the history and by aid of transillumination. A sarcoma situated in the posterior hemisphere or below the horizontal meridian will be hidden by detachment earlier than one anterior to the equator or in the upper part of the eye. Even while the tumor is small, more or less detachment of the retina usually exists below and, not infrequently, an extensive detachment may be produced by a very small sarcoma, in which case the latter may be overlooked. When the glaucomatous stage ensues the detachment is pushed forward against the lens, the retinal folds are crowded together and the broad surface furrows of simple detachment are transformed into narrow slits. Simple detachment usually occurs in myopic eyes, spontaneously or from slight injury. In any eye, detachment may be produced by traumatism, albuminuria, subretinal exudation or hemorrhage, contraction of retinal cicatrices, loss of support from diminution in the size of the vitreous and contraction of vitreous exudates. These causes must be considered and excluded before diagnosing a detachment as due to sarcoma of the choroid. Eye tension is not increased and may be diminished in simple detachment; it is elevated or at least normal in sarcomatous detachment. Sarcoma may be attended by discomfort, but simple detachment is painless. In simple detachment the retina may become temporarily replaced and a corresponding improvement in vision will occur, but sarcomatous detachment grows progressively worse. In simple detachment the loss of vision is rapid; in sarcoma gradual loss of vision usually precedes total detachment. Floating bodies in the vitreous are more common in myopic than in sarcomatous detachment. Detachment due to subretinal hemorrhage usually is accompanied by extensive bleeding in other parts of the retina. The picture of subhyaloid hemorrhage is too distinctive to be mistaken for tumor (Stergm. 33).

Hemorrhagic retinitis from venous thrombosis or other causes is not infrequently followed by inflammatory glaucoma. As in sarcoma of the choroid, the glaucoma is unilateral and preceded by painless loss of vision. The two conditions differ, however, in the following respects; in sarcoma vision usually disappears slowly and frequently the patient describes the gradual advance of darkness from one side, while in retinal hemorrhage vision is lost rapidly. In glaucoma due to sarcoma the anterior chamber is shallow, but in the early stage of glaucoma secondary

to retinal hemorrhage the anterior chamber usually is deep (see pages 151-2, 154).

Sarcoma usually blocks one or more of the vorticoses veins which empty the choroid, and a collateral flow is established through the anterior ciliary veins of the same side. Therefore, in a doubtful case, one or more greatly dilated anterior ciliary veins, limited to one side of the eye, are evidence in favor of sarcoma.

The form of the globe seldom is altered by sarcoma of the choroid until perforation is imminent, differing in this respect from conglomerate tubercle which tends to soften the overlying sclera, causing staphylomatous bulging. In one exceptional case which I microscopically examined,\* a large equatorial staphyloma existed over a sarcoma of the choroid. The cavity of the staphyloma was filled with disintegrated blood. The sarcoma had not invaded either the sclera or staphyloma, and the latter was produced by hemorrhage from either a vorticoses vein or from the sarcoma (Fig. 210).

Primary glaucoma almost always affects both eyes, though the disease may be incipient in one eye and advanced in the other; consequently, unilateral glaucoma should always suggest the presence of sarcoma. Glaucoma is attended by remissions and is more or less amenable to treatment, but in sarcomatous glaucoma the pain seldom remits until after perforation of the globe. Iridectomy, sclerotomy, etc., which relieve glaucoma, hasten the advance of sarcoma.

Sarcoma should be differentiated from metastatic carcinoma of the choroid, for the reason that sarcoma demands immediate enucleation, while in carcinoma of the choroid the fatal termination is precipitated by surgical interference, the average duration of life after enucleation being less than five weeks, although at the time the patient may appear in good health.† Therefore, in all cases of intraocular growth, search should be made for tumor elsewhere in the body, especially in the breast, and if a carcinoma is discovered the eye should not be enucleated except for relief of pain. Metastatic carcinoma of the choroid appears as a flat deposit at the posterior pole (Fig. 211), but when far advanced it may, like sarcoma, form a rounded protuberance in the vitreous. I examined microscopically one such case for Dr. P. C. Jameson. A woman, aged 38

\* From the service of Dr. J. Scott Wood, Brooklyn Eye and Ear Hospital.

† Oatman, *Am. Jour. Med. Sci.* March 1903.





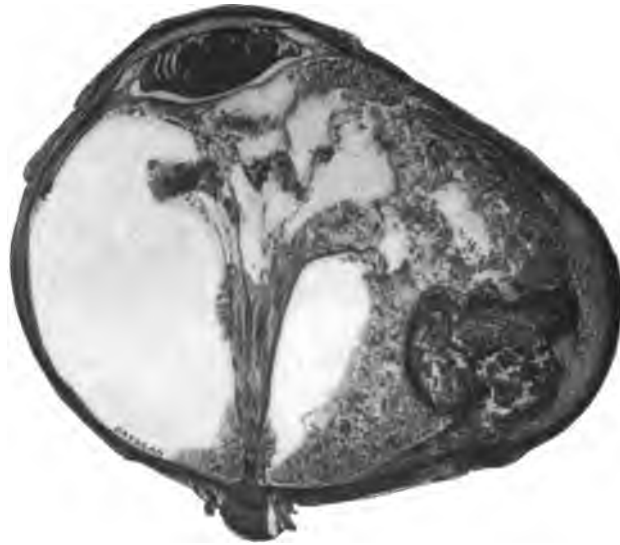


**Fig. 210.**

**Fig. 210—Sarcoma of the Choroid in Connection with a Large Equatorial Staphyloma, the latter caused by Hemorrhage and filled with Disintegrated Blood.**

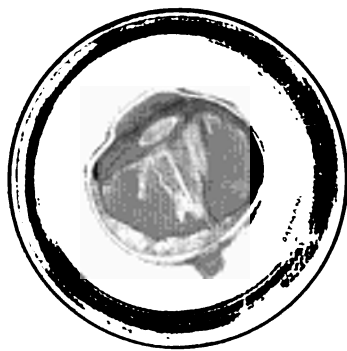


**Fig. 211.**

**Fig. 211—Metastatic Carcinoma of the Choroid, forming a Flat Tumor at the Posterior Pole.**



**Fig. 212.**

**Fig. 212—Metastatic Carcinoma of Choroid. There are two Deposits; one rounded and large at the Posterior Pole; the other flat, in the Ciliary Region.**

years, with carcinoma of the uterus, suffered from a metastatic carcinomatous deposit in the left eye.\* When first examined with the ophthalmoscope two deposits were seen: one, a large rounded tumor springing from the posterior pole, and a second mass in the ciliary region (Fig. 212).

#### SARCOMA OF THE CHOROID.

Always primary in the choroid.

May develop at any point.

Forms a rounded protuberance in the vitreous. Flat sarcoma (endothelioma) is extremely rare.

Usually exists for many months before destroying vision.

Detachment occurs late when tumor is centrally located (Griffiths).

The pain appears to be due solely to high eye tension.

If tumor is limited to choroid tension is not diminished.

Only one eye affected.

#### METASTATIC CARCINOMA OF THE CHOROID

Always secondary in the choroid.

Situated in posterior polar zone; usually on the temporal side of nerve.

Typical appearance is a flat, oval deposit with a central elevation of 1 or 2 mm., gradually sloping into the surrounding fundus. Its color is a dirty yellow with scattered pigment spots.

Vision is destroyed in from two to eight weeks after appearance of first eye symptoms.

Loss of vision is due to early and extensive detachment of retina.

Eye may be very painful with normal tension.

Tension is reported as increased in one-third, normal or diminished (?) in two-thirds of the cases.

In one-third of all cases both eyes contain a carcinomatous deposit.

\* The only recorded case of primary carcinoma of the uterus with metastasis in the choroid.

**STEREOGRAM 75. SARCOMA OF THE CHOROID WITH TOTAL DETACHMENT OF THE RETINA. Melanosarcoma.** Right eyeground of a man aged 47 years. Five months ago he first noticed a cloud in the upper field of this eye, which very gradually increased in size. Three weeks ago the eye rapidly became blind. *Present condition.* Right eye: The ciliary region is not reddened, but below a single dilated anterior ciliary vein appears on the sclera. The media are clear. Anterior chamber appears of normal depth. Eye tension, by digital palpation, is normal. Within the past day or two he has experienced a sense of discomfort in the eye. Vision is reduced to perception of light in the lower field and an absolute scotoma exists above. *Ophthalmoscope.* The retina is completely detached. The detachment is recognized as such by its folded, wavy surface, grey "shot-silk" color, regularly distributed and darkened retinal vessels and a trembling motion imparted by movement of the eye. The top of a dark brown nodular tumefaction which does not tremble when the eye is moved is seen in front of and below the detachment. Transillumination of the eye from a point far back and below, casts a shadow on the pupil. From other regions of the eye the pupil illuminates perfectly. The diagnosis is melanosarcoma of the choroid.

The history of this case indicates sarcoma. Development of the neoplasm was attended by slow, painless loss of vision. The abrupt blindness which ensued marks the time when detachment occurred.

The eye was enucleated and microscopic examination showed the tumor to be a well pigmented sarcoma of the choroid (Fig. 198).

#### *Bibliography.*

- Fuchs, d. Sarc. d. Uvealtract. Wien, 1882. Fuchs & Wintersteiner, B. d. o. G. 1900. Text Book.  
 Schieck, A. f. O., XLV, 2, 1898; XLVIII, w. 1899.

**GLIOMA OF THE RETINA.** Glioma of the retina is a malignant growth occurring only in infants and young children. It rarely, if ever, develops after the sixth year; even this is a late age for the first appearance of glioma and such cases require to be substantiated by authentic microscopic examination. Most gliomata are congenital, but owing to their slow progress they escape early observation. Usually, however, discovery occurs before the third year. Like sarcoma of the choroid, glioma of the retina passes through four stages:

(1) *Preglaucomatous stage*, in which it may be difficult or impossible to diagnosticate the condition;

(2) *Glaucomatous or inflammatory stage* which resembles the glaucomatous stage of sarcoma, except that in young children the sclera is more distensible than in adults and in glioma staphylomatous bulgings precede perforation;

(3) *Stage of extrabular extension.* Perforation is followed by lowered eye tension and relief from pain. If the opening is posterior, the neoplasm flourishes in the orbit and produces exophthalmus; if anterior, a granulating mass develops which has been termed fungus hematodes.

(4) *Stage of metastases and death.* Metastases occur early and show a preference for the lymphatic glands, including the preauricular, for the brain and for the cranial bones, especially the facial.

Glioma advances slowly by deposit of neighboring metastases and its course covers a period of several years, the victim finally dying from exhaustion or from metastatic deposits in vital parts, particularly in the brain. Death is inevitable unless the eye is enucleated before the stage of extraorbital extension. If the growth does not recur within three years after removal, the case may be regarded as cured. Glioma in the second eye appears to be an independent growth and not a metastasis or extension from the first eye; therefore, in bilateral glioma a cure may follow double enucleation. An eye enucleated for glioma should be microscopically examined in serial sections and, if tumor elements have passed into or beyond the sclera, the orbital contents should immediately be removed. Recurrence frequently is due to neglect of this precautionary measure. Infants born with manifest glioma frequently are affected in both eyes, while glioma which develops after birth is unilateral in about 90 per cent. of cases. When glioma develops in the second eye its evolution may be ophthalmoscopically observed. In the earliest state glioma may appear as several light colored spots which soon become elevated and develop an independent vascular supply. Usually, the first symptom of glioma to attract attention is a peculiar yellowish or pinkish white light-reflex that occasionally flashes through the pupil as from the eyes of certain animals; hence the condition has been known as amaurotic cat's eye. This pupillary reflex is not pathognomonic of glioma but occurs also in cases of detached retina saturated with inflammatory products, the so-called pseudoglioma.

STEREOGRAM 76. GLIOMA OF THE RETINA. Right fundus oculi of a boy, aged 2 years. Apparently in good health. Family history negative. Has had no eruptive fever or severe illness. The parents recently ob-

served a peculiar appearance of the right eye consisting of a whitish reflex from the pupil. Eye tension normal. Media clear. No iritic adhesions. The pupil responds promptly to direct action of light. By focal illumination a deep, pinkish white reflex is obtained through the pupil. With the ophthalmoscope the lower portion of the fundus is seen to be greatly elevated. On the nasal side below is a pinkish white tumefaction, covered with blood vessels which branch in an erratic manner and pursue anomalous courses. This mass has a firm appearance and does not tremble on movement of the eye, in fact, it presents the ophthalmoscopic picture of a neoplasm. On the temporal side and below the tumor are large, silver grey, bladder-like elevations, over which the retinal vessels pursue a normal course. This region trembles on sudden movement of the eye and, evidently, is a simple detachment of the retina. The pupil is not darkened on transillumination. The diagnosis is glioma with collateral detachment.

Enucleation was performed and microscopic examination showed the tumor to be a typical glioma retinae (Fig. 213). No extrabulbar extensions of the growth were discovered. Two years after enucleation there had been no recurrence.

**MORBID ANATOMY.** In glioma secondary deposits occur throughout the eye, a condition extremely rare in sarcoma. As a rule, glioma grows outward into the subretinal space (glioma exophytum); less frequently inward into the vitreous (glioma endophytum). The former type produces greater detachment of the retina than the latter. The optic nerve is early invaded by the tumor and converted into gliomatous tissue. The choroid is invaded both by metastasis and direct extension from the retina. Perforation usually occurs along the vessels which pass through the sclera and also along the optic nerve. Fresh glioma is pinkish white or light yellow in color and of soft consistence. From its gross resemblance to brain tissue it has been called medullary cancer. Retinal glioma is constructed of convoluted blood vessels the walls of which are composed of thickly packed tumor cells (Fig. 214). These cell-covered blood vessels are separated from each other by wide areas filled with dead cells which have perished from malnutrition. In the early stage of development glioma cells are arranged in radiate form around short, capillary tubules which frequently contain blood. Transverse microscopic sections of these tubules form the rosettes which, when present, are diagnostic of glioma (Fig. 215). In the later stages this peculiar structure is lost and the tumor presents the appearance of angiosarcoma. Even microscopically,

Fig. 213 — Glioma of the Retina.



Fig. 213.

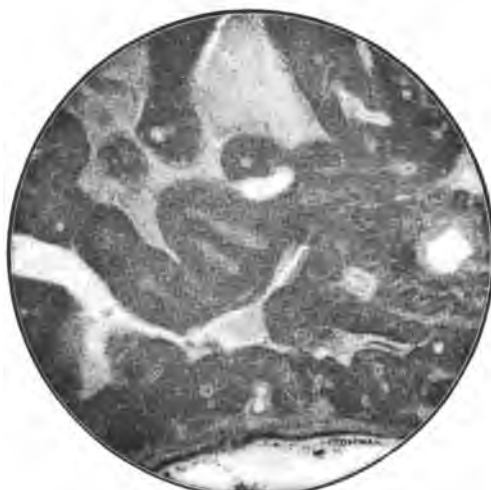


Fig. 214.

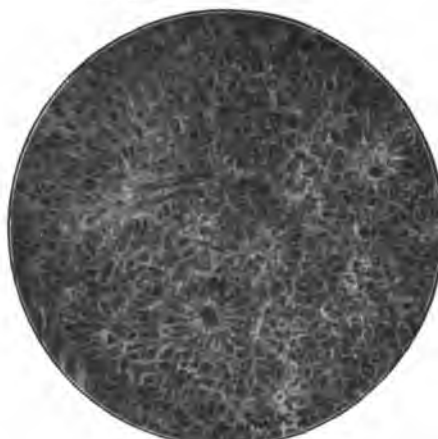


Fig. 215.

Fig. 214—Section of Retinal Glioma, showing the Convoluted Blood Vessels, the Walls of which are Filled with Tumor Cells.

Fig. 215—The same, showing the Tumor Cells arranged like Rosettes around the Walls of the Capillary Tubules.



pseudoglioma may present a resemblance to glioma sufficient to deceive the inexperienced. For example, I have examined a case of metastatic retinitis following epidemic cerebrospinal meningitis, in which the massed retinal tissue was thrown into folds which formed rosettes that might be mistaken for those of glioma. The variety of cells found in glioma of the retina indicates that some of them at least are common to all forms of chronic proliferative retinitis and have appeared in response to an irritant. The tumor cells are embryonic in type; they are of small size, round or cylindric in form and possess a large nucleus and scanty protoplasm. Ganglion and neuroglia spider cells can usually be demonstrated (Fig. 216). Glioma is strictly a nonpigmented tumor. If pigment is present it is either hematogenous or displaced from the retinal epithelium. The intercellular substance is scanty and consists largely of cell processes. The development of nutritive blood vessels in glioma does not keep pace with cellular proliferation; consequently, early and extensive necrosis and degeneration occur. The most common change is fatty degeneration followed by calcification. Less frequently hyaloid changes ensue. The metastases of glioma often histologically resemble small round-celled sarcoma. Glioma is said to arise from the nuclear layers of the retina. The name is based upon its supposed origin from the retinal neuroglia (Virchow). Other theories of origin are: (a) misplaced neuroglia and ganglion cells (Greeff); (b) misplaced rods and cones (Wintersteiner); embryonic retinal cells (Treacher Collins). The close structural resemblance of glioma to perithelioma (page 280) suggests a common vascular origin.

**CRYPTOGLIOMA.** Under the name of cryptoglioma Schöbl has described rare cases in which, although glioma is present, its symptoms are so obscured that a true diagnosis cannot be made. In these cases glioma develops in an eye but usually escapes observation; next, iridocyclitis develops, the globe softens and the fundus is hidden by exudates, cataract or detachment of the retina. At this period there is nothing except the age of the patient to suggest glioma. Later the glioma becomes active and progresses to a fatal termination. The cryptic character of these cases may be ascribed either to posterior perforation of the globe by the glioma, or to an iridocyclitis excited by the products of tumor necrosis. In another rare class of cases, glioma develops and then recedes without intercurrent iridocyclitis or softening of the eyeball. In course of time, however, the neoplasm reappears. Glioma possesses a great tendency to degenerate, and in these cases of recession it is probable that a large part of the tumor



dies and is removed by absorption. Cases of cryptoglioma are so rare as to exert but little influence upon differential diagnosis. Parsons found 15 alleged cases in literature. In but six of these was the microscopic examination at all satisfactory. Parsons says of all that "the gliomatous nature of cells in these shrunken eyes is not placed absolutely beyond cavil." Cases of supposed cryptoglioma require to be carefully differentiated from tuberculoma.

**PSEUDOGLIOMA.** The name "pseudoglioma" is applied to any non-malignant, intraocular formation in children which bears a clinical resemblance to glioma of the retina. Pseudoglioma usually is an inflammatory formation situated in either the retina, the choroid or both; less frequently in the vitreous. The majority of cases occur during the course of an infectious disease, particularly epidemic cerebrospinal meningitis, and are caused by bacterial metastasis. If a metastasis of pyogenic organisms lodges in the eye it is followed by suppuration of the globe (panophthalmitis). Pseudoglioma, however, follows a metastasis of nonpyogenic bacteria which excites plastic exudation rather than pus formation. Inasmuch as any of several intraocular parts may be the seat of a metastasis, the inflammation which results may be designated *metastatic endophthalmitis*. When both eyes are attacked the blindness which results immediately attracts attention, but a unilateral metastatic endophthalmitis occurring in the course of an acute fever usually escapes observation especially as the eye seldom exhibits external irritation, then or afterward. Intraocular infection occurs most frequently through penetrating wounds which either destroy the eye or leave the lens and cornea opaque. Occasionally, however, a wound may leave the media transparent and also result in formation of cicatricial tissue which, ophthalmoscopically, resembles a malignant neoplasm. In these traumatic cases the exudative process usually is more circumscribed than in metastatic endophthalmitis and it may be associated with simple detachment of the retina. A persistent fetal fibrovascular lens sheath has been mistaken for glioma. This malformation usually is associated with persistent hyaloid artery and undeveloped lens and capsule. It presents a shining white surface covered with blood vessels.

Various inflammatory conditions may result in a pseudoglioma. The most frequent is metastatic endophthalmitis with detachment of the retina. In these cases the posterior portion of the retina is gathered into a cord which extends from the optic nerve to the anterior part of the vitreous cavity where it expands to its attachment around the ora serrata,





**Fig. 216—Glioma of the Retina Showing Ganglion and Neuroglia Spider Cells.**

**Fig. 216.**

**Fig. 217—Umbrella-like Detachment of the Retina following Metastatic Endophthalmitis.**



**Fig. 217.**

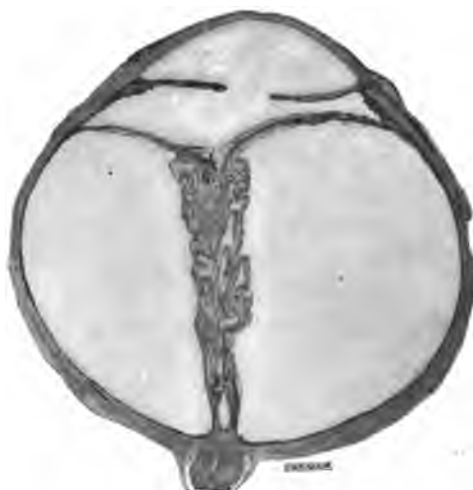


**Fig. 218.**

**Fig. 218—The same as 217. The Choroid is Not Detached in Either Specimen, best shown in Fig. 218.**

forming the so-called umbrella-form detachment (Figs. 217-218). In metastatic choroiditis the retina may not detach, but remain glued to the thickened choroid. Exudation from the ciliary body, especially after injury, may form a tumor-like mass in the vitreous which becomes encysted and may develop blood vessels. The majority of pseudogliomas which I have microscopically examined were cases of metastatic retinitis; less frequently metastatic choroiditis was found. In metastatic retinitis the contraction of cicatricial bands may drag the ciliary body from its bed even though there has been no cyclitis. If cyclitis develops, the eye softens.

As the result of disease or injury, exudate from the posterior portion of the ciliary body may be poured out into the post-lental region and organize into a permanent membrane on the anterior surface of the vitreous.

**STEREOGRAM 77. PSEUDOGLIOMA (Metastatic Endophthalmitis).** Left eyeground of a boy, seven years of age. His only severe sickness was from diphtheria, five years ago. Right eye, normal. His parents recently discovered that his left eye is blind. Since birth the left iris has been lighter in color than the right. No perception of light. The pupil reacts to a direct strong light, but is abnormally sluggish by weak illumination. Consensual pupillary reaction is normal. The ophthalmoscope shows the entire fundus to be unevenly elevated. Bluish white, nodular formations cover the disc and also the inferior and nasal portions of the eyeground. The upper outer quadrant of the fundus appears as an immovable, flat, pinkish white membrane. The entire surface of the eyeground is covered with blood vessels some of which are newly formed but the majority, from their method of distribution, are recognized as retinal. Many of the retinal vessels pierce the nodules, a peculiarity which suggests that the latter are composed of inflammatory exudate which has covered the vessels. The flat area extending upward and outward is supposed to be detached retina containing less exudate than other regions. The narrow, pointed, white formation in the lower border of the flat area is regarded as a band of newly formed connective tissue.

The diagnosis is metastatic endophthalmitis. Probably, the choroid was the seat of metastasis and the retina was fastened to the choroid by inflammatory exudates.

**DIAGNOSIS.** In the first stage, glioma presents no positive diagnostic sign; consequently, its recognition often is difficult or even impossible. Differentiation usually lies between glioma and a detached retina filled with inflammatory products (pseudoglioma). In these cases an accurate

decision is demanded. An eye containing a pseudoglioma is blind, but it is innocuous and may present a normal external appearance. Removal of such an eye disfigures the child and prevents proper development of the corresponding side of the face. On the other hand, failure to enucleate a gliomatous eye results in the patient's death. The history is important. In certain families several children perish from glioma retinae. Many solitary cases, however, occur. History of an infectious fever can usually be obtained in pseudoglioma. The presence of glioma is discovered before the third year in two-thirds of all cases, and the probabilities are strongly against its development beyond the age of five years. As our knowledge of glioma advances, late cases become more infrequent. Iritic adhesions and deposits on the lens are evidence in favor of pseudoglioma. Glioma nodules may develop on the iris in advanced cases, but are rare in the early stages when the diagnosis is in doubt. Glioma usually presents a clear anterior chamber, although hyphema and deposits of tumor debris somewhat resembling hypopyon have been reported. True hypopyon of a decided yellow color may form in metastatic endophthalmitis. Previous to the glaucomatous stage the pupillary reactions possess considerable diagnostic value. Thus, in glioma only a limited area of retina is destroyed and enough percipient elements remain to cause a prompt response of the pupil to light. Therefore, in glioma the pupil reacts well to direct light. In metastatic endophthalmitis, on the contrary, the entire retina may be converted into a fibrous membrane which retains few or no sensitive elements and the pupil will respond sluggishly or not at all to direct action of light, while consensual action is normal. Often, however, retinal tissue sufficient to produce pupillary response persists for a long time. Consequently, in pseudoglioma the pupil may or may not react to light, but an inactive pupil is evidence against the presence of glioma. As a rule, eye tension is normal or lowered in metastatic ophthalmitis and normal or heightened in glioma. In old metastatic endophthalmitis tension may rise from sudden recrudescence of the inflammatory process (Fig. 219) or from occlusion of the pupil in which cases the cause of the glaucoma is obvious. In nearly all doubtful cases, however, if tension is increased there is glioma and if lowered there is pseudoglioma. In soft eyes the possible existence of cryptoglioma should be borne in mind and the child kept under observation. The pinkish-white color of glioma may be duplicated in certain parts of an old inflammatory exudate, but other parts will exhibit either strong yellow, shining white or bluish colors that do





Fig. 219.

Fig. 219—Metastatic Endophthalmitis. Tension was Increased in this case on account of a Recrudescence of the Inflammatory Process.

Fig. 220—Massive, Circumscribed Exudation, due to Injury and Ophthalmoscopically Simulating Tumor.



Fig. 220.



Fig. 221.

Fig. 221—Post-lental Membranous Exudate (shown at P. in lower left of the figure).

not belong to glioma. The surface of glioma often presents discoloration due to hemorrhage, necrosis or calcification quite different in appearance from the bands and sheets of connective tissue seen in some cases of organized exudate. Adventitious blood vessels are nearly constant in glioma and not uncommon in pseudoglioma. Absence of all vessels on a tumor points to pseudoglioma. Inflammatory new formations may ensheath the vessels, an appearance which I have never seen in glioma. The appearance of the detached retina may throw some light on the nature of an intraocular tumor. Thus, in metastatic endophthalmitis the retina usually is totally detached and saturated with inflammatory material or converted into fibrous tissue which renders the membrane rigid and immovable on movement of the eye. In glioma, however, there may be extensive collateral simple detachment which trembles on movement; therefore, in a doubtful case the presence of a simple detachment with a tremulous surface points to the presence of glioma.

Massive, intraocular exudation following injury may present a puzzling ophthalmoscopic picture consisting of a circumscribed exudation (Fig. 220). In traumatic cases, however, evidence of an injury can usually be obtained.

Post-lental cyclitic membranes or exudates are best seen by focal illumination. They present a smooth, bluish-white surface which bears little resemblance to glioma (Fig. 221).

After metastatic choroiditis or uveitis, the vitreous cavity may be filled with a purulent fluid and the post-lental surface appears uniformly yellow. In advanced cases of glioma the preauricular glands enlarge and nodules may form on the head and facial bones.

Transillumination is less satisfactory in the diagnosis of glioma than in sarcoma. As a rule, pupillary illumination is but slightly dimmed by glioma. This is due to the following conditions: (a) glioma is a retinal growth and more or less space exists between tumor and sclera through which light reaches the pupil; (b) glioma contains no pigment; (c) the vascular interspaces are filled largely by degenerated tissue which does not obstruct light. A glioma may obstruct light if it has extensively invaded the choroid and especially if it also contains a large blood clot or has undergone extensive calcareous degeneration.

Practitioners unfamiliar with the appearance of intraocular growths sometimes mistake glioma for congenital cataract and delay treatment until the infant is older. This fatal error is avoided by the knowledge that congenital cataract lies immediately behind the pupil and possesses a



soft, bluish-white color, while glioma is deeply situated, far behind the pupil and has a whitish cast.

### *Bibliography.*

- Treacher Collins, *Researches*, London, 1896.  
 Greeff, *Lehrb. d. s. Path. Anat. (Orth.)*, II. Hälfte. I. Theil. 1903. Deutsch. m. Woch. XXII, 1896. D. Bau. u. d. Wesen d. Glioma, Deut. m. Woch. 1896, 327.  
 Parsons, *Path. of the Eye*, vol. II, 1905.  
 Schöbl, Norris and Oliver, III, 1898, 554.  
 Virchow, *Die Krankhaften Geschwulste*, Bd. II, 151.  
 Wintersteiner, *D. Neuroepithel. Ret.* 1897.

The following table is applicable only to the early stages of glioma of the retina, at which time the diagnosis is extremely difficult and often impossible:

|                            | GLIOMA.                                                                                                                                                                      | PSEUDOGLIOMA.                                                                                                                                             |
|----------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------|
| History.                   | May attack several children in a family. Not necessarily preceded by sickness or injury.                                                                                     | No family history of eye tumor. Preceded by some infectious fever or by injury.                                                                           |
| Age.                       | Usually discovered before the third year. Probabilities against its development after fifth year.                                                                            | May occur at any age.                                                                                                                                     |
| Anterior Chamber and Iris. | Anterior chamber usually clear. Iris is normal and free from adhesions.                                                                                                      | Iritic adhesions, atrophy of iris, hypopyon or deposits on lens or cornea, indicate an inflammatory process rather than glioma.                           |
| Pupillary Reactions.       | Early glioma affects only a limited area of retina; consequently, the eye retains good light perception and pupillary reactions are normal.                                  | Metastatic retinitis usually involves the entire retina, in which case all light perception <i>may be</i> lost and there is no direct pupillary reaction. |
| Tension.                   | Eye tension normal or increased.                                                                                                                                             | Eye tension frequently subnormal.                                                                                                                         |
| Detachment.                | Glioma often is accompanied by simple detachment of retina, which trembles on movement. Therefore, a tumor in infants, accompanied by simple detachment, probably is glioma. | In metastatic endophthalmitis the entire retina usually is infiltrated with exudates which render it rigid and immovable when the eye is moved.           |

**TUMORS OF THE CILIARY BODY. SARCOMA.** Sarcoma of the ciliary body usually is an extension of choroidal sarcoma. Primary sarcoma of the ciliary body rarely assumes the mushroom shape so frequent in sarcoma of the choroid, but forms a rounded tumor with a broad base which tends to grow forward into the iris and posterior chamber rather than backward into the choroid. As ciliary sarcoma advances it subluxates and deforms the lens but shows little tendency to invade it (Fig. 222).

The so-called ring sarcoma of the ciliary body is very rare (Fig. 201). It usually infiltrates and replaces the entire ciliary body before forming marked tumefactions. These growths are of relatively low malignancy and may exist many years before exciting suspicion as to their real character. The diagnosis of ring sarcoma is difficult. Transillumination is of no assistance unless the neoplasm has formed local tumefactions. In the late stage the iris may contain nodules or show increased pigmentation. Usually, however, it is only swollen and changed in color (Fuchs). If the ciliary processes are not destroyed glaucoma finally develops. Histologically, these growths are endotheliomata (pages 271-273).

Sarcomas which originate in the anterior portion of the choroid and extend to the ciliary body possess all the characteristics of choroidal sarcoma.

**STEREOGRAM 78. SARCOMA OF CHOROID AND CILIARY BODY.** Left fundus oculi of a man, 28 years of age. Vision in the left eye has recently fallen to counting fingers. No pain. Tension slightly minus. Externally, the eye is not reddened, but a single greatly dilated and tortuous anterior ciliary vein crosses the sclera on the nasal side. The ophthalmoscope reveals a very flat detachment of the retina in the macular region. At the anterior limits of the ophthalmoscopic field on the nasal side, the posterior edge of a light colored, slightly nodular tumor is seen. The surface of the mass is traversed by blood vessels which pursue a course the reverse of the retinal vessels, *i. e.*, from before backward. The posterior border of the tumor merges into loose folds of detached retina. The detachment presents a dark color owing to its position which prevents illumination by the choroidal reflex.\* Transillumination of the anterior nasal region corresponding to the intraocular mass, casts a shadow on the pupil. Diagnosis is sarcoma of the choroid and ciliary body.

\*In this stereogram the tumor is situated far forward in the eye, and when it is brought into focus with the ophthalmoscope, all details in the posterior part of the fundus are lost.

The eye was enucleated and found to contain a poorly pigmented sarcoma (Fig. 223).

Conglomerate tubercle of the ciliary body may simulate sarcoma. It differs, however, in its tendency to invade all structures that lie in its path including the iris, lens (Fig. 224) and sclera. In ciliary tubercle secondary deposits usually form in the iris and excite a low grade, plastic inflammation. In other cases conglomerate tubercle of the ciliary body may attain large size and perforate the globe without exciting the slightest local reaction (Fig. 225).

**DIAGNOSIS.** Tuberculosis of the ciliary body usually occurs at an earlier age than sarcoma. In suspicious cases a tuberculin injection may be employed for diagnostic purposes and a positive reaction in the eye is proof that the process is tubercular.

Metastatic carcinoma may appear in the ciliary body. In a case which I examined microscopically for Dr. J. Duncan, the tumor cells extended backward infiltrating the choroid, thus following the line of least resistance (Fig. 226).

The diagnosis of metastatic ciliary carcinoma rests upon the knowledge of carcinoma elsewhere in the body.

Glioma has been known to originate in the pars ciliaris retinae, although in the great majority of cases ciliary glioma is secondary to a primary growth in the retina. Glioma is the malignant tumor of infancy and does not enter into the differential diagnosis of intraocular growths in adults.

A case of metastatic hypernephroma of the ciliary body has been reported by Burton Chance\* who permitted me to examine microscopic slides of the growth (Fig. 227). The diagnosis depends upon the known presence of hypernephroma.

Cysts may form in the pars ciliaris retinae of degenerated eyes by separation of its layers (Fig. 171). They occur also in eyes which appear otherwise normal, especially in old people (Fig. 228). They should be carefully differentiated from melanosis by means of transillumination.

Sometimes the uveal pigment on the ciliary processes forms large melanotic masses which might be mistaken for malignant growths (Fig. 229). The benign melanotic masses which I have seen present a scintillating surface when viewed with the ophthalmoscope. Owing to their separation from the sclera they do not cast a shadow in transillumination.

I once microscopically examined an eye enucleated for supposed sarcoma of the ciliary body which on section was found to contain no sar-

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\* Chance, Jour. Am. Med. Assn., Feb. 9, 1907, 475.

**Fig. 222—Sarcoma of the Ciliary Body. The Tumor has Deformed and Luxated the Lens.**



**Fig. 222.**



**Fig. 223—Poorly Pigmented Sarcoma of the Choroid and Ciliary Body.**

**Fig. 223.**

**Fig. 224—Conglomerate Tubercle of the Ciliary Body Invading the Lens.**



**Fig. 224.**



Fig. 225 — Con-  
glomerate Tubercle  
of the Ciliary Body  
Perforating the  
Sclera.



Fig. 225.

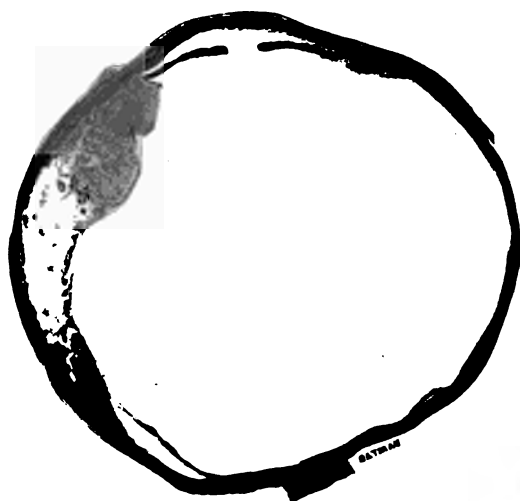


Fig. 226—Metastatic Carcinoma of  
the Ciliary Body.

Fig. 227—Metastatic Hydrone-  
phroma of the Ciliary Body.



Fig. 227.





Fig. 228.

Fig. 228—Cyst of the Pars Ciliaris Retinæ.



Fig. 229.

Fig. 229—Masses of Uveal Pigment Simulating Malignant Growths of the Ciliary Body.







Fig. 230—Telangiectasis of the Ciliary Body. Large Cystic Spaces also exist beneath the Epithelial Layer



Fig. 230.

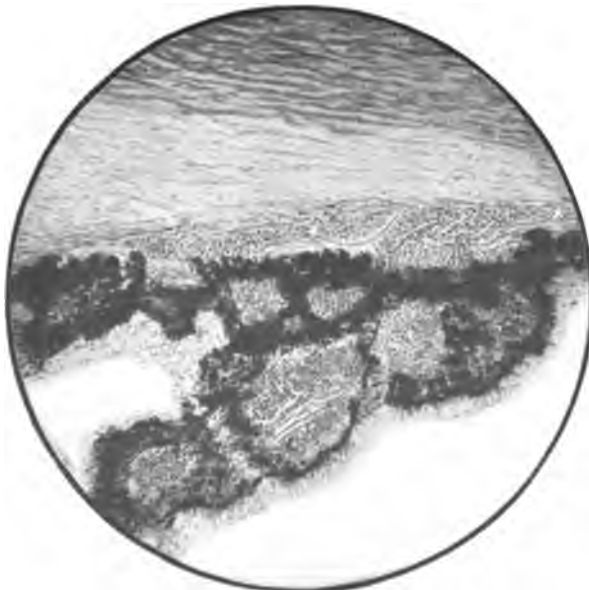


Fig. 231.

Fig. 231—Same as Fig. 230. The Mesh of Blood Vessels is covered by Dense Masses of Uveal Pigment, forming Melanomata.

coma but an angiomatous formation, probably congenital. The clinical appearance was remarkably deceptive. The subject was a man, 46 years of age. Myopia of 7. D. in each eye. Recent loss of vision in the left eye. Ophthalmoscope showed extensive, temporal detachment of the retina including the macula (Fig. 120). Immediately in front of the detachment and connected with the ciliary body, was a nodular black mass, containing blood vessels. This case occurred before transillumination was employed in eye diagnosis. The eye was enucleated. Microscopic examination showed that the tumor seen with the ophthalmoscope was formed by the long ciliary artery which, on entering the ciliary processes, immediately broke up into a telangiectatic mesh of vessels densely covered with uveal pigment which in places formed melanomata (Figs. 230-231). In addition, several large cystic spaces existed beneath the epithelial layer. The detachment of the retina, which caused the loss of vision and was supposed to be due to the ciliary tumor, proved to be simple in its nature and such as is common in myopic eyes.

**TUMORS OF THE ORBIT AND OPTIC NERVE.** Extraocular tumors may induce changes in the fundus of the eye consisting of optic neuritis, optic atrophy and secondary hypermetropia from pressure on the ocular walls.

**ORBITAL TUMORS** may be malignant or benign.

**DERMOID CYSTS** are the most common of all growths originating in the orbit. They are always congenital, although they may be so small at birth as to escape observation. The great majority are situated in front of the eye and do not cause exophthalmus unless, as in some cases, they extend deeply into the orbit.

**FLUID IN TENON'S CAPSULE** or on the tendons of muscles may simulate cysts.

**HYDATID CYSTS** usually occur in the young, two-thirds of all cases occurring between the ages of 11 and 21 years (Neisser).

**INFLAMMATORY TUMORS** and bulging inward of the orbital walls are due to disease or tumors in the adjacent nasal sinuses.

**GRANULATIONS** and **POLYPI** in the orbit are connected with bone disease, usually originating in adjoining nasal cavities.

**PRIMARY EXOSTOSES** of the orbit usually spring from the upper and inner angle, although they may grow from any portion of the bony wall. They are composed of an extremely dense, bony shell containing a center of spongy bone (ivory exostosis); rarely, they are partly cartilaginous. When situated in the sphenoidal fissure, the optic nerve is

compressed and atrophy results. Osteoma may invade the orbit from without. Such cases should be carefully differentiated, for the reason that primary tumors situated superficially are easily removed, whereas it is highly dangerous to operate upon those which have extended into the orbit from adjoining cavities.

SIMPLE ANGIOMA (telangiectasis) and CAVERNOUS ANGIOMA both occur in the orbit. Simple angiomata are congenital growths, situated in the lids, whence they extend into the orbit. They are not, as a rule, encapsulated. Cavernous angiomata develop in the orbit. They possess a thick fibrous capsule, lined with endothelium and are crossed by trabeculae, the structure resembling erectile tissue. They may contain phleboliths. Bruit and pulsation are not present in these tumors.

LYMPHIANGIOMATA seldom occur in the orbit.

LIPOMATA are reported as occurring in the orbit but such diagnoses must be accepted with caution inasmuch as the amount of adipose tissue normally present varies widely within physiologic limits.

LYMPHOMATA occur in the orbit under two forms, (1) leukemic and pseudoleukemic deposits, both of which are associated with disease of the blood and blood forming organs; (2) round celled growths resembling sarcoma but apparently nonmalignant.

NEUROMATA, *i. e.*, growths consisting of newly-formed nerve elements, and neurofibromata have been observed in the orbit. Plexiform neuroma (Elephantiasis Neuromatodes, Cirroid Neuroma) occurs in the orbit very rarely.

FIBROMATA may grow from the periosteum, sheaths of tendons and dural sheath of the optic nerve. In diagnosing fibroma of the orbit, the tendency of other growths, especially angiomata, to undergo fibrous hyperplasia must be borne in mind.

CARCINOMA does not occur primarily in the orbit but invasion may occur from the skin, conjunctiva, lachrymal gland and from metastasis in the choroid.

SARCOMA may originate in the orbit although in the great majority of cases it is an extension or metastasis from the choroid.

GLIOMA OF THE ORBIT is always a secondary deposit from glioma of the retina.

CYLINDROMA (endothelioma) forms encapsulated tumors in the orbit. They are of very low malignancy.

The prognosis after operation in orbital tumors is far more favorable when the tumor is encapsulated than when it is diffuse.



Fig. 232—Orbital Tumor causing Localized Hypermetropia by Pressure upon, and Thickening of, the Sclera. S, sclera; T, tumor.



Fig. 232.



Fig. 233.

Fig. 233—Tumor of the Optic Nerve.

Fig. 234—The so-called Glioma of the Optic Nerve, showing its development from the Connective Tissue Septa.

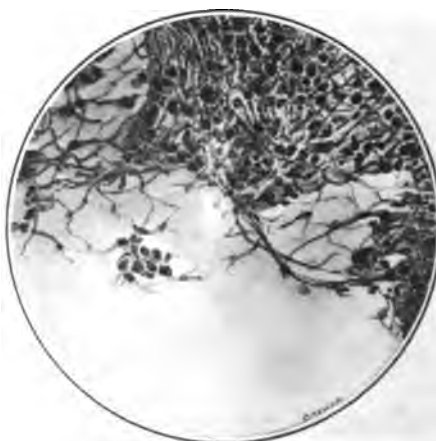


Fig. 234.

LACHRYMAL GLAND TUMORS are very rare. Frequently, they are mixed growths. Carcinoma, sarcoma, myxoma, myxadenoma, myxosarcoma, enchondroma, osteochondroma, osteoma, fibroma, adenoma, adenoangioma, cylindroma, conglomerate tubercle and hydatid cysts have been reported as occurring in the lachrymal gland.

Protusion of the eyeball (exophthalmus) is the most prominent symptom of orbital tumor. If situated without the muscle cone, the eye is pushed to one side according to the situation of the growth. Unlike tumors of the optic nerve, orbital growths do not produce early blindness; consequently, diplopia usually is present. A tumefaction at the apex of the orbit, however, may destroy sight from pressure on the optic nerve. Pressure of an orbital growth on the eyeball may push the walls inward or cause the sclera to thicken, in which case the ophthalmoscope will show a localized hypermetropia (Fig. 232).

TUMORS OF THE OPTIC NERVE. Tumors of the optic nerve are divided into two classes, intradural and extradural. The latter are very rare and develop around the nerve outside the sheaths. Strictly speaking, only those growths which develop within the dural sheath should be considered as primary tumors of the nerve.

Intradural tumors of the optic nerve mostly begin in early life or are congenital. Out of 85 cases collected by Byers, 52 appeared before the 25th year. They are of slow development and low malignancy. As a rule, they extend backward from the entrance of the central vessels, leaving 10 mm. to 12 mm. of the optic end of the nerve free. They may also extend forward and press against the globe.

Optic nerve tumors vary in size from a slight enlargement to the size of an egg (Fig. 233). In a large proportion of cases they are connected with an intracranial extension which is not removed with the tumor, and the continued growth of which ultimately destroys life. They are usually elastic or of soft consistency and may contain large cystic spaces. Histologically, they belong to the connective tissue group. The most characteristic nerve tumor is myxoma or myxosarcoma. Endotheliomata also are common. When these tumors contain chalky concretions they are called psamomata. Fibroma and fibrosarcoma are rare. Greeff says the so-called glioma of the nerve develops from the connective tissue septa of the nerve, the neuroglia taking no part in their formation (Fig. 234). Were the tissue of these neoplasms proliferating neuroglia they would react to selective glia stains, which they fail to do. The term "neuroma" when applied to these tumors also is a misnomer, because



the optic nerve is brain tissue, which does not proliferate. Greeff applies Cohnheim's theory to the development of intradural tumors. He believes that when the central vessels enter the fetal cleft in the nerve, a certain number of embryonic cells are carried along, which persist and ultimately proliferate, forming a tumor of the nerve.

The exophthalmus of optic nerve tumor is characteristic. The eye is thrust directly forwards with little or no lateral displacement. Motility is not disturbed until exophthalmus is extreme. The eyeball cannot be pushed back into its normal position. Blindness occurs early, often before the exophthalmus attracts attention, in which event the patient has no diplopia. Usually, there is first choked disc followed by postneuritic atrophy. The central vessels in the nerve are destroyed but this occurs so gradually that the retinal circulation is continued by the new-formed vessels in the tumor without inducing either ischemia or stasis. The development of new vessels around the ocular end of the nerve may explain why circulation persists in the retina after removal of nerve and tumor. When exophthalmus is extreme the lids cannot cover and protect the cornea which becomes dry, desiccated and, finally, ulcerated (*keratitis e lagophthalmo*). In this condition the lower half of the cornea suffers, chiefly because this portion is uncovered, especially during sleep when the lids are at rest and the cornea directed upward. Exceptionally, pressure of the tumor may advance the posterior pole of the eye, thus producing a secondary hypermetropia.

STEREOGRAM 79. OPTIC ATROPHY AND SECONDARY HYPERMETROPIA FROM TUMOR OF THE OPTIC NERVE.\* Left eyeground of apparently healthy girl, 14 years of age. Left exophthalmus first attracted attention two months ago, at which time she had transient diplopia. At present, the eye is blind. Exophthalmus is marked, the displacement being directly forward. The eye cannot be restored by pressure to its normal position. No pain. Motility is unimpaired. The cornea and media are transparent. The optic disc presents the dead white color of postneuritic atrophy. Retinal vessels normal. The entire posterior pole is elevated above other parts of the fundus. The center of this area has an elevation of 2 mm. (6. D.); the periphery slopes gradually away to the level of the normal fundus. The projection does not resemble detached retina but gives one the impression that the posterior walls of the globe are pushed inward. It is crossed by several horizontal light streaks, presumably caused by inequalities in the surface.

---

\* Patient was seen through courtesy of Dr. Frank Van Fleet who reported case in Medical Record, June 27, 1908.

A tumor of the optic nerve was discovered and removed by the Krönlein operation.

**DIAGNOSIS.** A tumor of the nerve produces anterior displacement of the eye; a tumor of the orbit produces lateral displacement. In nerve tumors motility of the eye is good; in orbital tumors it is defective. The eye is blind in tumor of the nerve, but some vision is retained in orbital growths; therefore, diplopia exists in the latter condition but not in tumor of the nerve except, possibly, in the early stage. Age is an important factor in differentiating these conditions, inasmuch as about 40 per cent of intradural nerve tumors develop before the tenth year. In exophthalmus due to solid tumor the eye cannot be pushed backward into its normal position, differing in this respect from exophthalmic goitre and telangiectasis of the orbit in which the displaced eye can temporarily be reduced by pressure. The secondary hypermetropia presented by the above case is unusual in tumor of the nerve.

*Bibliography.*

- Byers. Stud. from R. Victoria Hosp. Montreal, 1901.  
Neisser, Die Echinokockenkrank., Berlin, 1877.  
Greef, Orth's Lehrb. d. spez. Path. Anat. II 1, Theil, p. 450.

1

## INDEX.

### A

|                                                            | PAGE |
|------------------------------------------------------------|------|
| Albinism of the Eyeground, acquired (Stereogram 26).....   | 55   |
| Albinism of the Eyeground, congenital (Stereogram 15)..... | 33   |
| Albuminuric Retinitis (see Retinitis) .....                | 81   |
| Amaurosis, toxic .....                                     | 208  |
| Amaurosis, uremic .....                                    | 87   |
| Amaurotic Family Idiocy (Stereogram 57).....               | 200  |
| Amblyopia, alcoholic .....                                 | 209  |
| Amblyopia, from iodoform .....                             | 210  |
| Amblyopia, from quinine .....                              | 210  |
| Amblyopia, from salicylic acid .....                       | 210  |
| Amblyopia, from tobacco .....                              | 209  |
| Amblyopia, from wood alcohol .....                         | 208  |
| Amblyopia, toxic .....                                     | 208  |
| Atrophia Gyrata Choroidæ et Retinæ .....                   | 181  |
| Atrophy of the Choroid (see Choroid) .....                 | 55   |
| Atrophy of Optic Nerve (see Optic Nerve) .....             | 231  |

### B

|                   |     |
|-------------------|-----|
| Buphthalmus ..... | 264 |
|-------------------|-----|

### C

|                                           |         |
|-------------------------------------------|---------|
| Canal of Cloquet (Stereogram 14) .....    | 32      |
| Choked Disc (Stereograms 63-64) .....     | 220     |
| Choked Disc, causes of .....              | 221     |
| Choked Disc, in brain tumor .....         | 222-224 |
| Choked Disc, theories of .....            | 220     |
| Choriocapillaris .....                    | 12      |
| Choroid .....                             | 11      |
| Choroid, affections of .....              | 37      |
| Choroid, atrophy of (Stereogram 26) ..... | 55      |
| Choroid, congenital anomalies of .....    | 20      |
| Choroid, conglomerate tubercle of .....   | 51      |

|                                                                                | PAGE   |
|--------------------------------------------------------------------------------|--------|
| Choroid, miliary tuberculosis of .....                                         | 51     |
| Choroid, rupture of (Stereogram 28) .....                                      | 59     |
| Choroid, sarcoma of (Stereogram 74-75) .....                                   | 270    |
| Choroid, sclerosis of .....                                                    | 54     |
| Choroid, tuberculosis of (Stereogram 23) .....                                 | 48     |
| Choroid, tumors of .....                                                       | 270    |
| Choroid, vascular degeneration of .....                                        | 54     |
| Choroidal Ring .....                                                           | 18     |
| Choroiditis, central (Stereograms 19-21) .....                                 | 42     |
| Choroiditis, disseminated (Stereograms 17-18) .....                            | 37     |
| Choroiditis, disseminated, differentiation .....                               | 39     |
| Choroiditis, disseminated, stage of cicatrization (Stereogram 20) .            | 43     |
| Choroiditis, disseminated, stage of repair .....                               | 44     |
| Choroiditis, disseminated, symptoms of .....                                   | 41     |
| Choroiditis, exudative .....                                                   | 37     |
| Choroiditis, metastatic .....                                                  | 215    |
| Choroiditis, syphilitic (Stereogram 19) .....                                  | 42     |
| Choroiditis, suppurative .....                                                 | 37-215 |
| Chorioretinitis, diffuse, atrophic stage (Stereogram 24) .....                 | 52     |
| Chorioretinitis Pigmentosa (Stereogram 22) .....                               | 46     |
| Chorioretinitis Pigmentosa, differentiation from retinitis pigmentosa          | 48     |
| Ciliary Body, tuberculosis of .....                                            | 288    |
| Ciliary Body, tumors of .....                                                  | 287    |
| Cilio-retinal Arteries .....                                                   | 15     |
| Circle of Zinn .....                                                           | 16     |
| Coloboma, Fuchs' (Stereogram 10) .....                                         | 26     |
| Coloboma of the Choroid (Stereogram 6) .....                                   | 20     |
| Coloboma of the Choroid, differentiation from old cicatrices .....             | 21     |
| Coloboma of the Choroid, pathogenesis .....                                    | 22     |
| Coloboma of Choroid and Optic Nerve (Stereogram 7) .....                       | 21     |
| Coloboma of the Macula (Stereogram 8) .....                                    | 23     |
| Coloboma of the Optic Nerve and Sheath (Stereogram 9) .....                    | 24     |
| Coloboma of the Optic Nerve and Sheath, differentiation from<br>glaucoma ..... | 26     |
| Colloid Excrescences (Stereogram 27) .....                                     | 57     |
| Commotio Retinæ (Stereogram 59) .....                                          | 205    |
| Crescent, inferior (Stereogram 10) .....                                       | 26     |
| Crick Dots (Stereogram 16) .....                                               | 35     |
| Cyanosis Retinæ (Stereogram 58) .....                                          | 204    |

## D

|                                                      | PAGE |
|------------------------------------------------------|------|
| Degenerations of the Retina .....                    | 179  |
| Detachment of the Retina (see Retina) .....          | 159  |
| Diabetic Retinitis (see Retina) .....                | 94   |
| Diaphanoscopy .....                                  | 7    |
| Direct Method of Ophthalmoscopy (Stereogram 1) ..... | 3    |
| Disseminated Choroiditis (see Choroid) .....         | 37   |

## E

|                                                                   |       |
|-------------------------------------------------------------------|-------|
| Embolism of the Central Artery (see Retinal Vessels) .....        | 105   |
| Endogenous Ophthalmitis .....                                     | 215   |
| Examination of the Eye .....                                      | 1     |
| Examination of the Eye, by determination of the visual fields.... | 6     |
| Examination of the Eye, by tonometry .....                        | 9, 10 |
| Examination of the Eye, by transillumination .....                | 7     |
| Examination of the Eye, with oblique illumination .....           | 5     |
| Examination of the Eye, with the ophthalmoscope .....             | 1     |

## F

|                                                       |    |
|-------------------------------------------------------|----|
| Fuchs' Coloboma (Stereogram 10) .....                 | 26 |
| Fundus Oculi, normal (Stereogram 2) .....             | 11 |
| Fundus Oculi, senile changes in (Stereogram 27) ..... | 57 |
| Fundus Oculi, negroid (Stereogram 3) .....            | 14 |

## G

|                                                       |             |
|-------------------------------------------------------|-------------|
| Glaucoma .....                                        | 251         |
| Glaucoma, absolutum .....                             | 253         |
| Glaucoma, acute .....                                 | 252         |
| Glaucoma, causes of .....                             | 257 et seq. |
| Glaucoma, chronic .....                               | 254         |
| Glaucoma, degenerativum .....                         | 254         |
| Glaucoma, fulminans .....                             | 254         |
| Glaucoma, hemorrhagic (Stereogram 73) .....           | 255-265     |
| Glaucoma, infantile (buphthalmus) .....               | 264         |
| Glaucoma, ophthalmic picture of (Stereogram 72) ..... | 255         |
| Glaucoma, pathogenesis of .....                       | 257         |
| Glaucoma, pathology of .....                          | 262         |

|                                               | PAGE    |
|-----------------------------------------------|---------|
| Glaucoma, primary .....                       | 251     |
| Glaucoma, secondary .....                     | 260     |
| Glaucoma, simple (Stereogram 72) .....        | 254-256 |
| Glioma of the Retina (Stereogram 76) .....    | 278     |
| Glioma of the Retina, diagnosis of .....      | 283-286 |
| Glioma of the Retina, morbid anatomy of ..... | 283     |
| Gunn's Dots (Stereogram 16) .....             | 35      |

## H

|                                                         |         |
|---------------------------------------------------------|---------|
| Hemorrhage, intravitreal, recurrent .....               | 77      |
| Hemorrhage, intravitreal, recurrent, causes of .....    | 78      |
| Hemorrhage, preretinal (Stereogram 33) .....            | 76      |
| Hemorrhage, retinal .....                               | 71      |
| Hemorrhage, retinal, etiology of .....                  | 73      |
| Hemorrhage, subhyaloid (Stereogram 33) .....            | 76      |
| Hemorrhagic Albuminuric Retinitis (Stereogram 35) ..... | 90      |
| Hemorrhagic Diabetic Retinitis (Stereogram 38) .....    | 95      |
| Hemorrhagic Leukemic Retinitis (Stereogram 39) .....    | 98      |
| Hemorrhagic Neuroretinitis (Stereogram 47) .....        | 139-142 |
| Hereditary Macular Anomaly .....                        | 197     |
| Hereditary Optic Atrophy .....                          | 235     |
| Hyaloid Artery, persistent (Stereogram 14) .....        | 32      |

## I

|                                               |    |
|-----------------------------------------------|----|
| Indirect Method of Ophthalmoscopy .....       | 4  |
| Inferior Optic Crescent (Stereogram 10) ..... | 26 |
| Intraocular Pressure .....                    | 9  |
| Intravitreal Hemorrhage .....                 | 77 |

## L

|                                       |    |
|---------------------------------------|----|
| Lamina Vitrea .....                   | 12 |
| Leukemic Retinitis (see Retina) ..... | 96 |

## M

|                                                                 |     |
|-----------------------------------------------------------------|-----|
| Macula Lutea .....                                              | 13  |
| Macula, Coloboma of (Stereogram 8) .....                        | 23  |
| Maculo-cerebral Degeneration (familial) (Stereograms 55-56) ... | 191 |

|                                                          | PAGE        |
|----------------------------------------------------------|-------------|
| Maculo-cerebral Degeneration, macular type .....         | 192-194     |
| Maculo-cerebral Degeneration, maculo-cerebral type ..... | 191-192-193 |
| Maculo-cerebral Degeneration, differentiation .....      | 197         |
| Maculo-cerebral Degeneration, etiology .....             | 196         |
| Maculo-cerebral Degeneration, pathology .....            | 195         |
| Medullated Nerve Fibers (Stereogram 11) .....            | 27          |
| Metastatic Ophthalmitis (Stereogram 77) .....            | 215         |
| Myopia .....                                             | 244         |
| Myopia, anatomical changes in .....                      | 246         |
| Myopia, etiology of .....                                | 244         |
| Myopia, non-progressive (Stereogram 70) .....            | 246         |
| Myopia, progressive (Stereogram 71) .....                | 248         |
| Myopia, varieties of .....                               | 244         |

## N

|                                                                      |             |
|----------------------------------------------------------------------|-------------|
| Neuritic Optic Atrophy (Stereogram 69) .....                         | 236         |
| Neuritis, Optic (Stereograms 31, 35, 36, 65, 66) .....               | 220         |
| Neuritis, Optic, in cerebral sinus thrombosis (Stereogram. 66) ..... | 228         |
| Neuritis, Optic, in disease of nasal sinuses (Stereogram 65) .....   | 225         |
| Neuritis, Optic, with albuminuric retinitis (Stereogram 36) .....    | 91          |
| Neuro-retinitis, hemorrhagic .....                                   | 138-139-142 |

## O

|                                                                             |     |
|-----------------------------------------------------------------------------|-----|
| Oblique, or Focal Illumination .....                                        | 5   |
| Ophthalmoscope .....                                                        | 1   |
| Opaque Nerve Fibers (Stereogram 11) .....                                   | 27  |
| Optic Atrophy .....                                                         | 231 |
| Optic Atrophy, following occlusion of central vessels (Stereogram 44) ..... | 133 |
| Optic Atrophy, following injury of orbit .....                              | 236 |
| Optic Atrophy, hereditary .....                                             | 235 |
| Optic Atrophy, in disease of the pituitary body .....                       | 235 |
| Optic Atrophy, in lateral sclerosis of the cord .....                       | 235 |
| Optic Atrophy, in multiple sclerosis (Stereogram 68) .....                  | 233 |
| Optic Atrophy, in paresis .....                                             | 235 |
| Optic Atrophy, in syringomyelia .....                                       | 235 |
| Optic Atrophy, in tabes (Stereogram 67) .....                               | 231 |
| Optic Atrophy, post-neuritic (Stereogram 69) .....                          | 236 |
| Optic Atrophy, retinitic (Stereograms 22-53) .....                          | 180 |



|                                                        | PAGE |
|--------------------------------------------------------|------|
| Optic Atrophy, simple .....                            | 231  |
| Optic Crescent, inferior (Stereogram 10) .....         | 26   |
| Optic Nerve, anatomy of .....                          | 18   |
| Optic Nerve, atrophy of .....                          | 231  |
| Optic Nerve, compression of, in arteriosclerosis ..... | 239  |
| Optic Nerve, tumors of .....                           | 291  |

## P

|                                                             |       |
|-------------------------------------------------------------|-------|
| Papilledema (Stereograms 63-64) .....                       | 220   |
| Physiologic Excavation of the Disc (Stereograms 5-40) ..... | 18    |
| Postneuritic Optic Atrophy (Stereogram 69) .....            | 236   |
| Post-thrombotic Changes in the Fundus (Stereogram 49) ..... | 156   |
| Prepapillary Retinal Artery (Stereogram 12) .....           | 29    |
| Preretinal Hemorrhage (Stereogram 33) .....                 | 76    |
| Pseudoglioma (Stereogram 77) .....                          | 282   |
| Pseudoneuritis (Stereogram 13) .....                        | 30    |
| Pulsation of Retinal Vessels .....                          | 16    |
| Pulsation of Retinal Vessels, in arteriosclerosis .....     | 62-64 |
| Punctate Condition of the Fundus (Stereogram 16) .....      | 35    |

## Q

|                         |     |
|-------------------------|-----|
| Quinine Amblyopia ..... | 210 |
|-------------------------|-----|

## R

|                                                        |         |
|--------------------------------------------------------|---------|
| Retina, normal .....                                   | 13      |
| Retina, angioid streaks in .....                       | 213     |
| Retina, cyanosis of (Stereogram 58) .....              | 204     |
| Retina, detachment of (Stereograms 50-51-52) .....     | 159     |
| Retina, detachment of, diagnosis .....                 | 164     |
| Retina, detachment of, etiology .....                  | 168     |
| Retina, detachment of, incipient (Stereogram 50) ..... | 160     |
| Retina, detachment of, old (Stereogram 52) .....       | 166     |
| Retina, detachment of, prognosis .....                 | 168     |
| Retina, detachment of, recent (Stereogram 51) .....    | 163     |
| Retina, detachment of, serous .....                    | 159     |
| Retina, detachment of, symptoms .....                  | 161-167 |
| Retina, glioma of (Stereogram 76) .....                | 278-279 |

|                                                                            | PAGE        |
|----------------------------------------------------------------------------|-------------|
| Retina, miscellaneous affections of .....                                  | 204         |
| Retina, re-attachment of .....                                             | 165         |
| Retina, traumatic edema of (Stereogram 59) .....                           | 205         |
| Retinal Hemorrhage .....                                                   | 71          |
| Retinal Hemorrhage, etiology of .....                                      | 73          |
| Retinal Reflexes (Stereogram 3) .....                                      | 14          |
| Retinal Vessels, normal (Stereogram 4) .....                               | 15          |
| Retinal Vessels, closure of .....                                          | 105         |
| Retinal Vessels, closure of central artery (Stereog. 41, 42, 43, 45) ..... | 105         |
| Retinal Vessels, closure of central artery, anatomic characters....        | 126         |
| Retinal Vessels, closure of central artery, clinical history .....         | 132         |
| Retinal Vessels, closure of central artery, etiology .....                 | 113         |
| Retinal Vessels, closure of central vein (Stereograms 46-47-48) ..         | 138         |
| Retinal Vessels, closure of central vein, etiology .....                   | 140         |
| Retinal Vessels, closure of central vein, clinical history .....           | 150         |
| Retinal Vessels, closure of central vein, complications .....              | 151         |
| Retinal Vessels, closure of central artery and vein .....                  | 156         |
| Retinal Vessels, sclerosis of (Stereograms 29-30) .....                    | 62          |
| Retinal Vessels, sclerosis of, diagnosis .....                             | 66          |
| Retinal Vessels, sclerosis of, etiology .....                              | 63          |
| Retinal Vessels, sclerosis of, morbid anatomy .....                        | 68          |
| Retinal Vessels, sclerosis of, with hemorrhages (Stereogram 32) ..         | 70          |
| Retinal Vessels, sclerosis of, with optic neuritis (Stereogram 31) ..      | 69          |
| Retinitis, Albuminuric (Stereograms 35, 36, 37) .....                      | 81          |
| Retinitis, Albuminuric, acute .....                                        | 86          |
| Retinitis, Albuminuric, differentiation from choroiditis .....             | 91-92       |
| Retinitis, Albuminuric, with hemorrhages .....                             | 90          |
| Retinitis, Angiopathic (Stereogram 34) .....                               | 80          |
| Retinitis, Circinata (Stereogram 40) .....                                 | 101-104     |
| Retinitis, Diabetic (Stereogram 38) .....                                  | 94          |
| Retinitis, Leukemic (Stereogram 39) .....                                  | 96          |
| Retinitis, Metastatic .....                                                | 215         |
| Retinitis, Pigmentosa (Stereogram 53) .....                                | 179         |
| Retinitis, Pigmentosa sine Pigmento .....                                  | 181         |
| Retinitis, Proliferans (Stereogram 61) .....                               | 211         |
| Retinitis, Punctata Albescens (Stereogram 54) .....                        | 181-189-190 |
| Retinitis, Septic .....                                                    | 52-217      |
| Retinitis, Striata (Stereogram 62) .....                                   | 213         |

|                                                               | PAGE |
|---------------------------------------------------------------|------|
| Retinitis, with Pernicious Anemia .....                       | 98   |
| Retinitis, with Syphilis .....                                | 100  |
| Retinitis, with Wood Alcohol, Poisoning (Stereogram 60) ..... | 208  |

## S

|                                                              |        |
|--------------------------------------------------------------|--------|
| Sarcoma of the Choroid (Stereograms 74-75-78) .....          | 270    |
| Sarcoma of the Ciliary Body (Stereogram 78) .....            | 287    |
| Sclera .....                                                 | 11     |
| Sclerosis of the Choroid (Stereogram 25) .....               | 54     |
| Sclerosis of the Retinal Vessels (see Retinal Vessels) ..... | 62     |
| Senile Changes in the Fundus (see Fundus) .....              | 57     |
| Septic Retinitis .....                                       | 52-217 |
| Subhyaloid Hemorrhage (Stereogram 33) .....                  | 76     |
| Suprachoroidea .....                                         | 11     |
| Staphyloma, ciliary .....                                    | 263    |
| Staphyloma, intercalary .....                                | 263    |
| Staphyloma, posterior, in myopia .....                       | 247    |
| Staphyloma, verum .....                                      | 247    |

## T

|                                                                 |     |
|-----------------------------------------------------------------|-----|
| Tabetic Atrophy of the Optic Nerve (Stereogram 67) .....        | 231 |
| Tay's Disease .....                                             | 200 |
| Thrombosis of Central Retinal Vein (Stereograms 46-47-48) ..... | 138 |
| Tobacco Amblyopia .....                                         | 209 |
| Tonometer .....                                                 | 10  |
| Tonometry, digital .....                                        | 9   |
| Tonometry, instrumental .....                                   | 10  |
| Toxic Amblyopia and Amaurosis .....                             | 208 |
| Transillumination .....                                         | 7   |
| Tuberculosis of the Choroid (see Choroid) .....                 | 48  |
| Tuberculosis, of the Ciliary Body (see Ciliary Body) .....      | 288 |
| Tumors, intraocular and intraorbital .....                      | 270 |
| Tumors, of the ciliary body .....                               | 287 |
| Tumors, of the choroid .....                                    | 270 |
| Tumors, of the optic nerve .....                                | 291 |
| Tumors, of the optic nerve causing hyperopia (Stereogram 79) .. | 292 |
| Tumors, of the orbit .....                                      | 289 |

INDEX.

303

U

|                               |                          |
|-------------------------------|--------------------------|
| <b>Uremic Amaurosis</b> ..... | <b>PAGE</b><br><b>87</b> |
|-------------------------------|--------------------------|

V

|                                                                   |           |
|-------------------------------------------------------------------|-----------|
| <b>Vascular Degeneration of the Choroid (Stereogram 25)</b> ..... | <b>54</b> |
| <b>Vitreous Hemorrhage, recurrent</b> .....                       | <b>77</b> |
| <b>Vitreous Hemorrhage, recurrent, causes of</b> .....            | <b>78</b> |
| <b>Vitreous Hemorrhage, recurrent, prognosis</b> .....            | <b>79</b> |

Z

|                              |           |
|------------------------------|-----------|
| <b>Zinn, Circle of</b> ..... | <b>16</b> |
|------------------------------|-----------|







